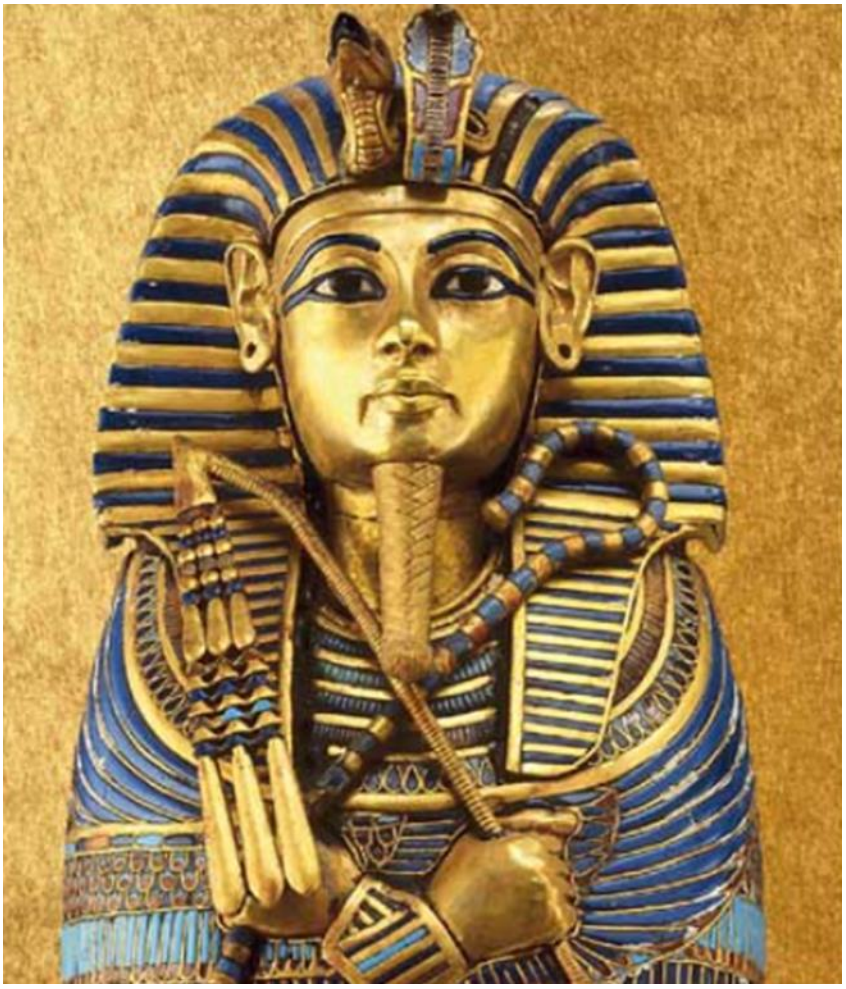
NEUROIMAGING



Compiled by:

Dr. M. Samy, MD

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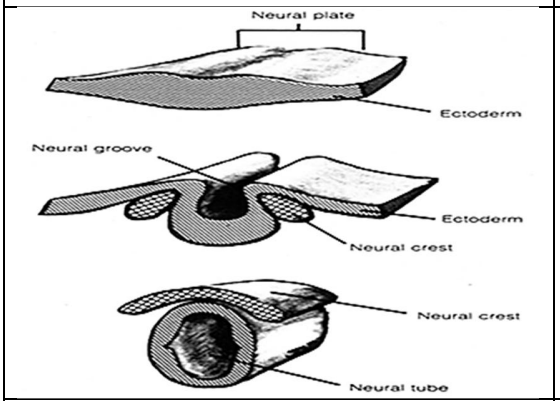
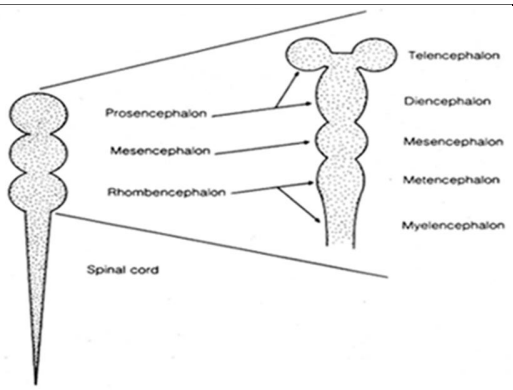
Congenital CNS Anomalies

<table>
<tr><th colspan="2">Stages Of Embryological Development</th></tr>
<tr><td>Stage 1: Dorsal Induction</td><td>Stage 2: Ventral Induction</td></tr>
<tr><td>2 – 4 w</td><td>5 – 10 w</td></tr>
<tr><td>Formation and Closure of the Neural Tube</td><td>Formation of Brain Segments and Face</td></tr>
<tr><td><u>Failure:</u>
• Anencephaly
• Cephalocele
• Chiari</td><td><u>Failure:</u>
• Holoprosencephalies
• Corpus callosum agenesis
• Dandy Walker
• Facial anomalies</td></tr>
<tr><td> 
</td><td> 
</td></tr>
<tr><td>Stage 3: Migration and Histogenesis</td><td>Stage 4: Myelination</td></tr>
<tr><td>2 – 5 m</td><td>5 – 10 m</td></tr>
<tr><td>a. Neural proliferation
b. Neural differentiation
c. Neural migration</td><td></td></tr>
<tr><td><u>Disorders:</u>
• Heterotopia
• Agyria-pachyria, polymicrogyria
• Teratomas
• Phakomatosis</td><td>Failure:
• Developmental delay
• Dysmyelinating disease</td></tr>
</table>

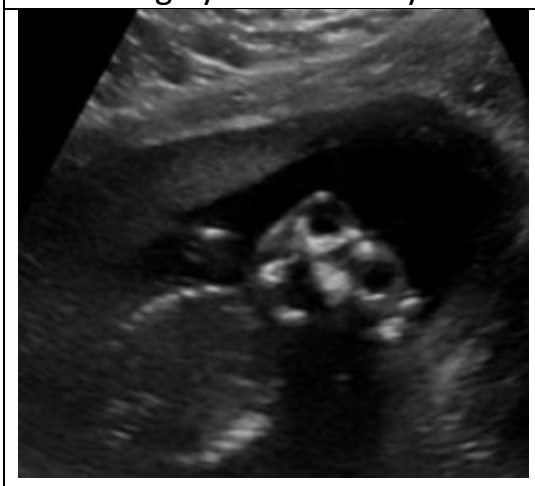
Specific Anomalies Presented in This Discussion include: (after van der Knaap (71)	Timing of Gestational Defect
1. Dorsal Induction	
A. Chiari malformations	4 weeks
B. Encephaloceles	4 weeks
2. Ventral Induction	
A. Holoprosencephaly	5–6 weeks
B. Septooptic dysplasia	6–7 weeks
C. Dandy-Walker malformation	7–10 weeks
3. Neuronal Proliferation and Histogenesis	
A. Neurofibromatosis	5 weeks–6 months
B. Tuberous sclerosis	5 weeks–6 months
C. Primary hydranencephaly	3 months or later
4. Migration	
A. Schizencephaly	2 months
B. Agyria and pachygyria	3 months
C. Gray matter heterotopias	5 months
D. Dysgenesis of corpus callosum	2–5 months

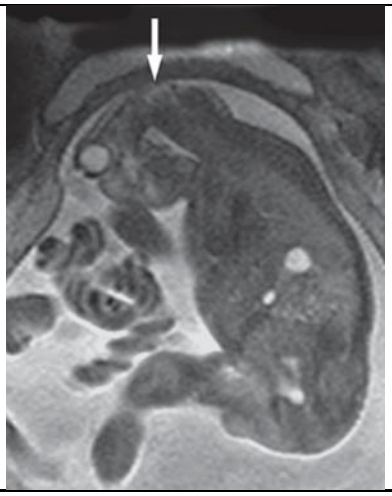
Stage I Failure

1) Anencephaly

- Failure of the brain and skull development.
- Most severe anomaly.
- Ultrasound diagnosis as early as 20weeks.
- Polyhydramnios
- High maternal serum alpha fetoprotein.
- Death.

- A "frog eye" or "mickey mouse"





2) Cephalocele

- Intracranial contents herniate through a congenital defect in the skull and dura.

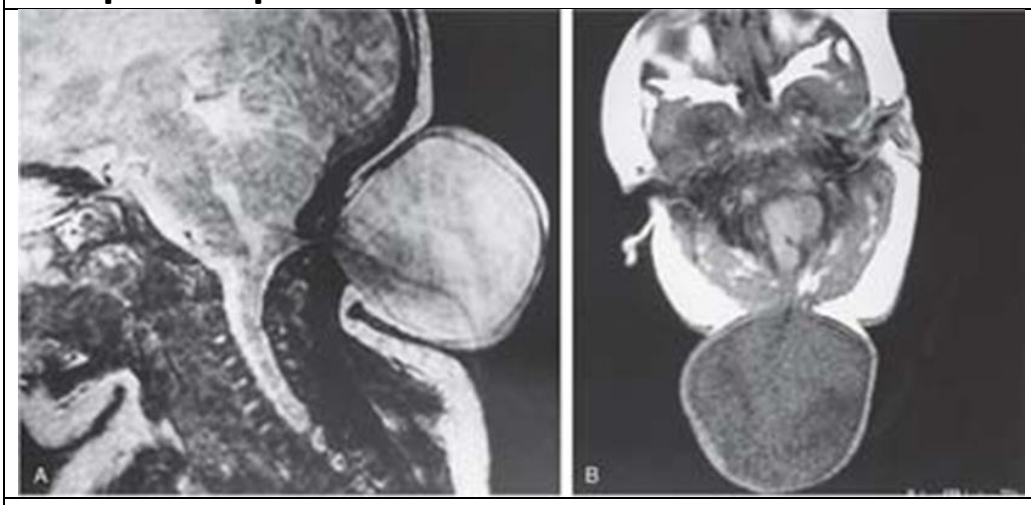
Contents :

- Meningocele (herniation of meninges with CSF)
- Meningoencephalocele (herniation of brain and meninges).
- Atretic cephalocele (herniation of dura & degenerated brain)
- Gliocele: (herniation of glial lined cyst containing CSF).

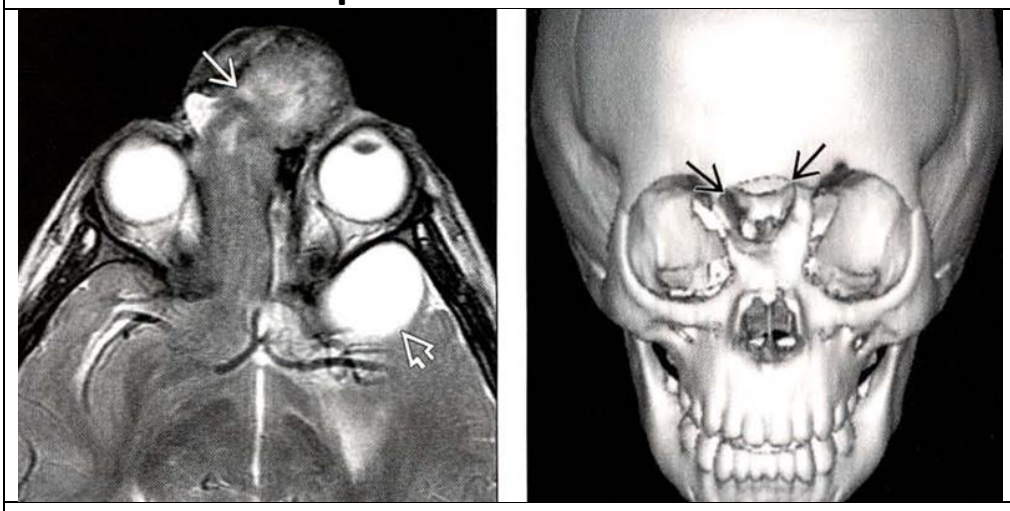
Location:

- Occipital
- Frontal
- Parietal
- Skull base

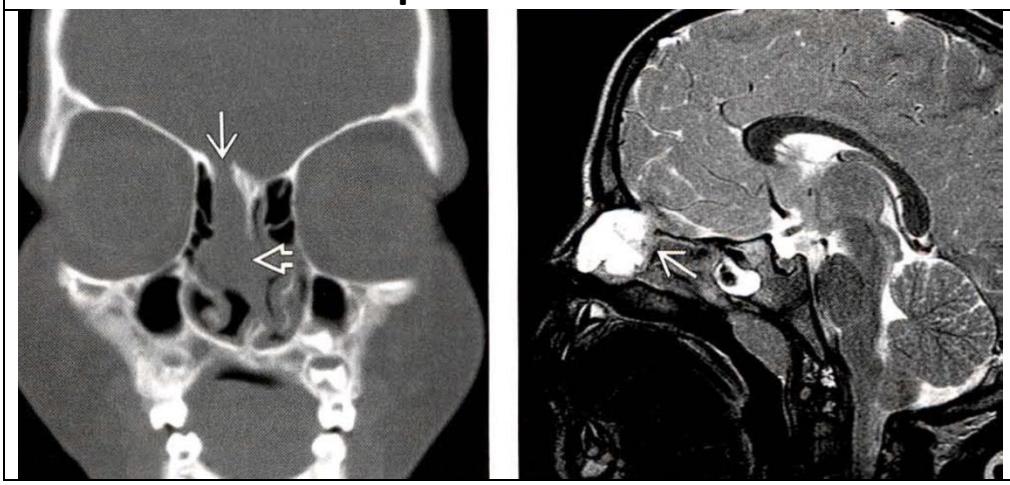
Occipital Cephalocele



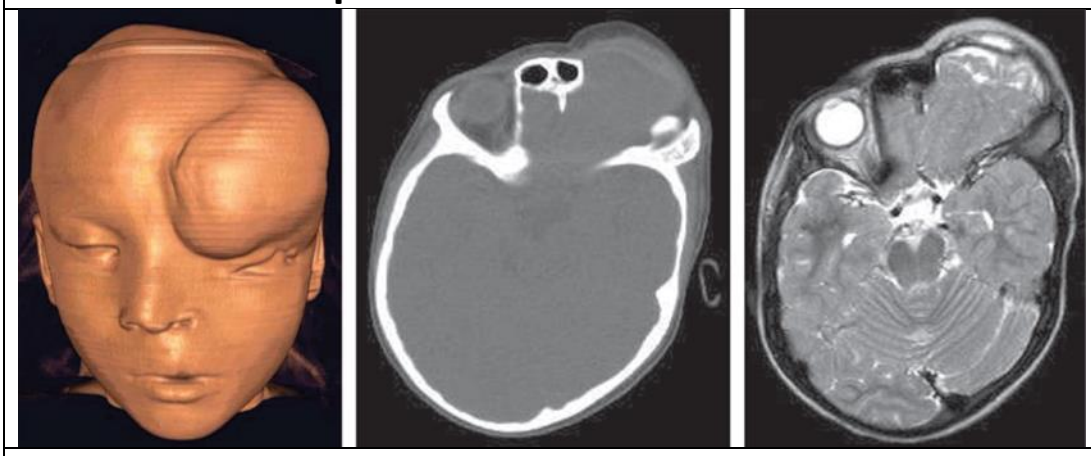
Fronto-Nasal Cephalocele



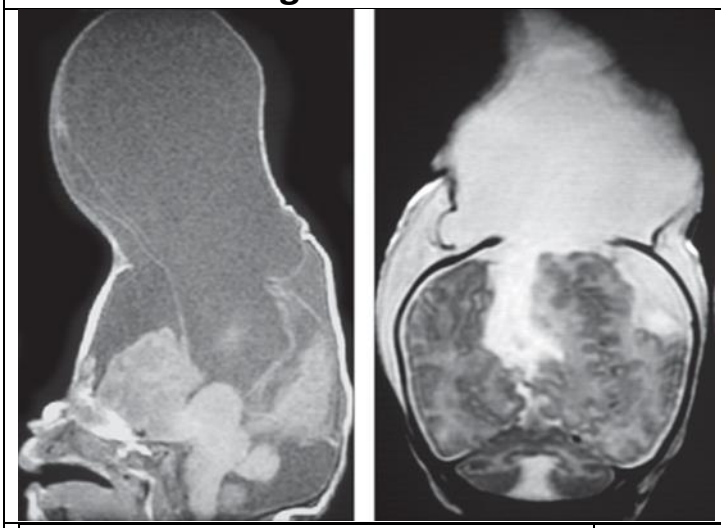
Naso-Ethmoidal Cephalocele



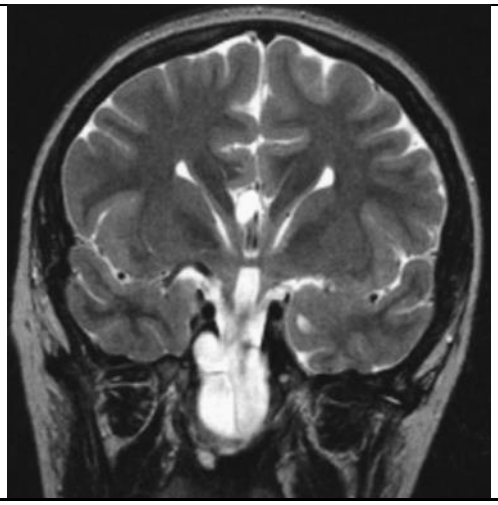
Naso-Orbital Cephalocele:



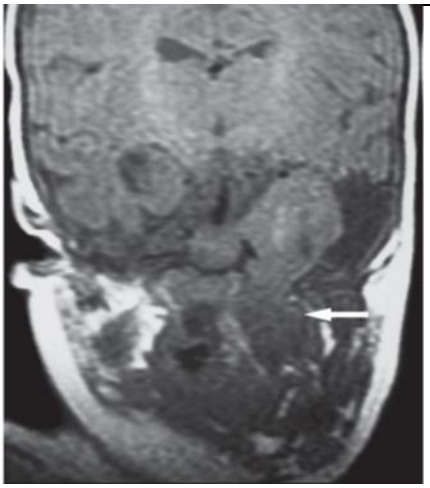
Parietal Meningocele



Trans-Sphenoidal Cephalocele

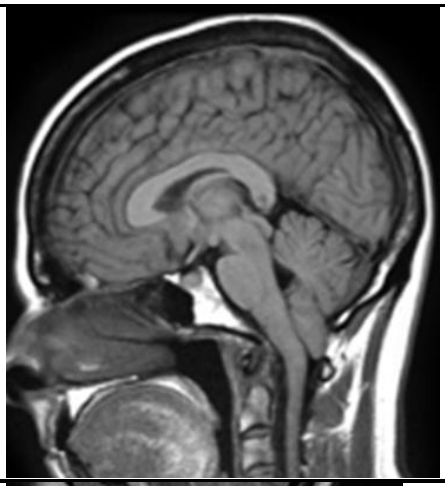


Skull Base Cephalocele



3) Chiari I

- Caudal protrusion of "peg-shaped" tonsils below foramen magnum (FM)
- Tonsils with peg/triangular shape + obliteration of surrounding CSF
  >6mm in young
  >3mm in adults



Associations:-

- Syrinx
- Klippel-Feil syndrome
- Sprengel deformity
- Platybasia
- NF1

DD: Acquired Tonsillar Herniation

- Basilar invagination

"Pull from below"

- Intracranial hypotension
- Lumbar puncture or (LP) shunt
- Spontaneous intracranial hypotension

"Push from above"

- Chronic ventriculo-peritoneal shunt (thick skull, premature sutural fusion, lumbar arachnoidal adhesions)
- Tonsillar herniation 2ry to ICP, mass effect or tumor

4) Arnold Chiari II

A- Small Posterior fossa

- Below: "Cascade" or "waterfall" of tissue down, behind medulla
- Above: "Towering" cerebellum protrudes up through incisura,
- Anterior: cerebellar hemispheres wrapping around the brainstem
- 4th ventricle: Elongated, "straw-like" without posterior point (fastigium)
- Low-lying torcular
- Obliterated basal cisterns
- Beaked tectum.
- Medullary kink.
- Concave clivus.

B- Supratentorial:

- Lateral V: Pointed anterior horns + Colpocephaly
- Large massa intermedia, high-riding vein.
- CC dysgenesis
- Occiptal lobe polymicrogyria (Stenogyria)
- Fenestrated/hypoplastic falx
- Interdigitated gyri

C- SPINE:

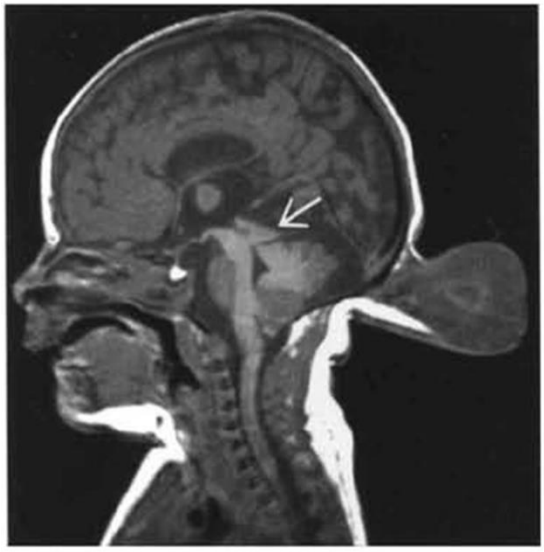
- Open dysraphism 100% (lumbar> >cervical)
- Hydrosyringomyelia 20-90%
- Posterior arch C1 anomalies 66%
- Diastematomyelia 5%

Associated Anomalies:

- Myelomeningocele (88-100%)
- Dysgenesis of corpus callosum (80-90%)
- Obstructive hydrocephalus following closure of myelomeningocele (50-98%)
- Syringohydromyelia (50-90%)
- Aqueductal stenosis (70%)
- Absence of septum pellucidum (40%)
- Contracted narrow gyria (stenogyria; 50%)
- Heterotopias
- Diastematomyelia
- Segmentation anomalies (<10%), incomplete C1 arch
- Malrotation of the posterior arches of C1 and C2
- Low-lying often-tethered conus medullaris below lumbar nerve L2

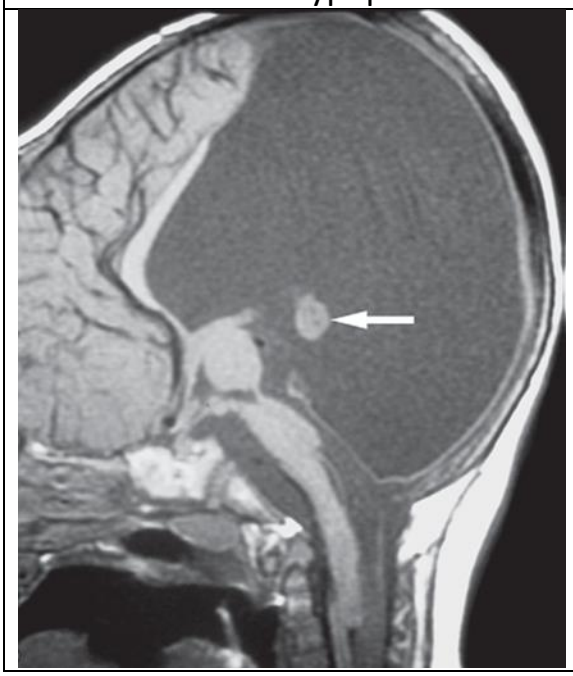
5) Chiari III

Chiari II malformation + High cervical/occipital meningoencephalocele



6) Chiari IV

Severe cerebellar hypoplasia.

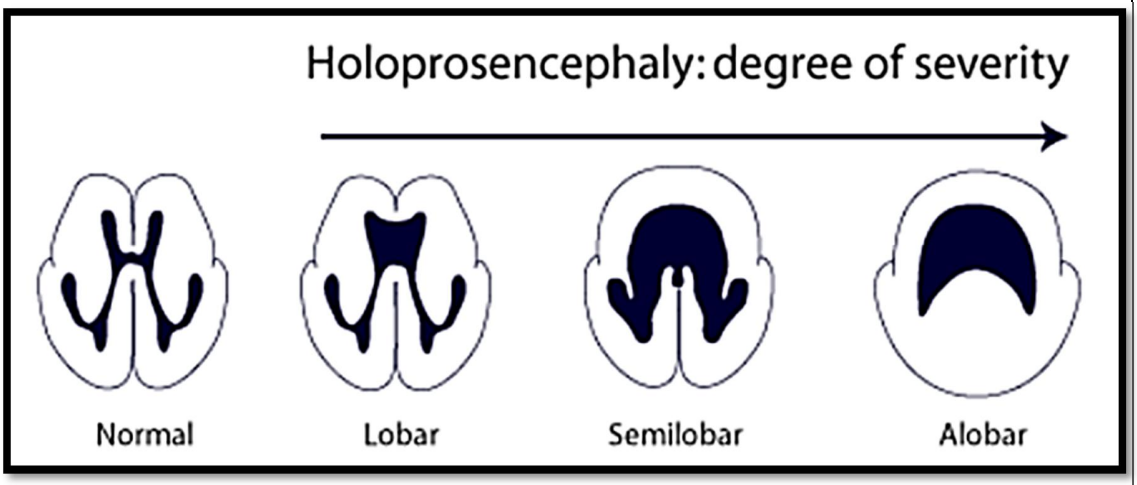


Stage II Failure

1) Holoprosencephaly (HPE)

An incomplete or absent division of the embryonic forebrain (prosencephalon) into distinct lateral cerebral hemispheres in 5th gestational week

HPE Types	HPE Variants:
Alobar form Semilobar form Lobar form	Middle Interhemispheric Variant (MIH) = Syntelencephaly. Septo-Optic dysplasia (SOD)

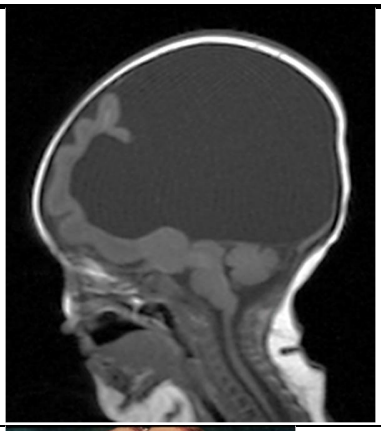


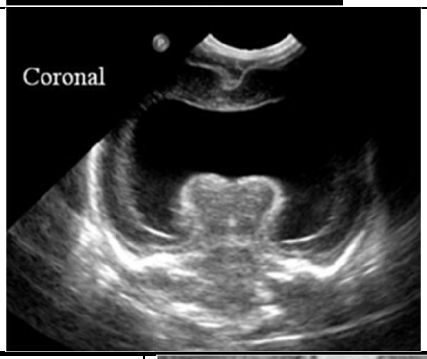
THREE TYPES OF HOLOPROSENCEPHALY

	Alobar	Semilobar	Lobar
Interhemispheric fissure and falx	Absent	Present posteriorly	Present*
Lateral ventricles	U-shaped monoventricle	Partially fused anteriorly	Near normal
Third ventricle	Absent	Rudimentary	Near normal
Cerebral hemisphere	One brain	Partial formation	Near normal
Thalamus	Fused	Variable fuison	Near normal
Facial anomalies	Severe	Less severe	None or mild
Septum pellucidum	Absent	Absent	Absent

A- Alobar Holoprosencephaly

Absent all midline structures	Fusion
Absent falx Absent interhemispheric fissure Absent corpus callosum Absent septum pellucidum Absent fornix. Absent 3rd ventricle	Fused cerebral hemispheres Fused Thalami Single mono-ventricle



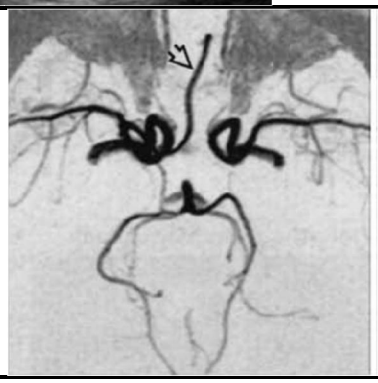


MRA

- Azygous or absent ACA

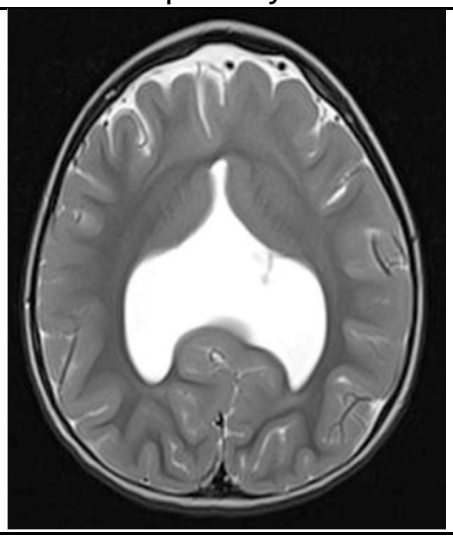
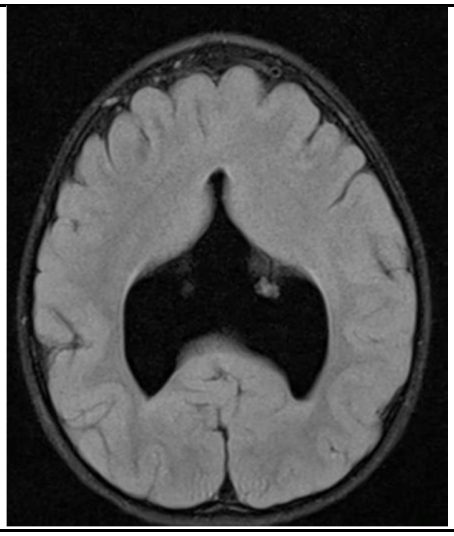
MRV

- Absent superior sagittal, inferior sagittal and straight sinuses
- Cortical veins and deep veins drain directly to torcular



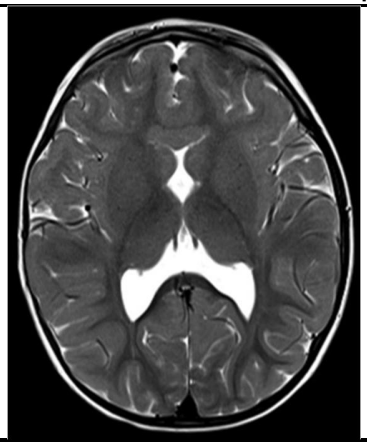
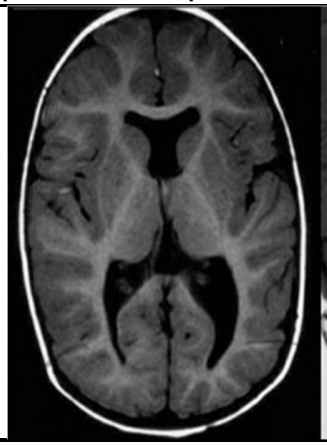
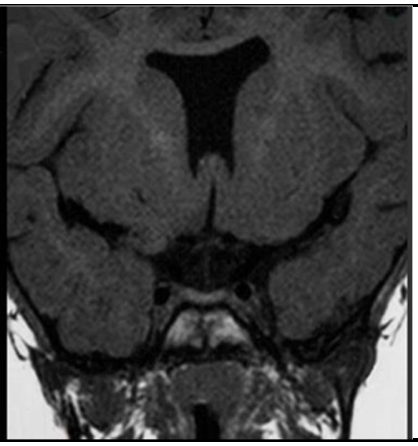
B- Semi Lobar HPE

- Frontal brain is fused
- Partial separation of the posterior occipital and temporal lobes.
- Thalami partially fused

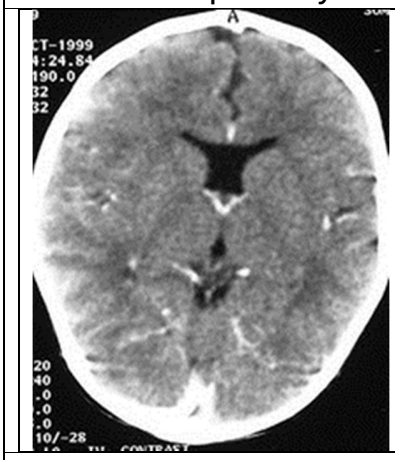
C- Lobar HPE

- Absent septum pellucidum
- Fusion of only anterior inferior frontal lobes.
- Otherwise the brain appears to be quite normal.

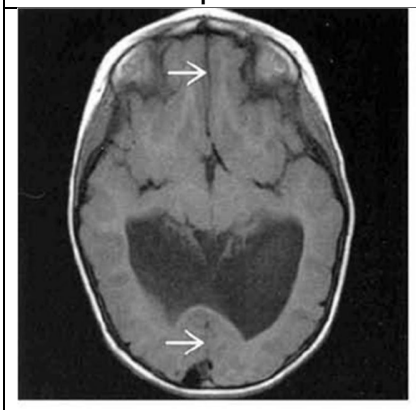
D- Septal Optic Dysplasia:

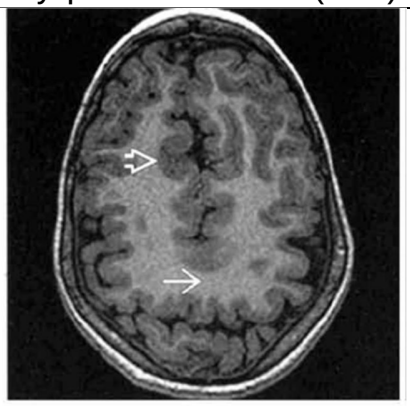
- Most mild form
- No septum pellucidum
- Flat roof of lateral ventricles,
- Box like frontal horns "pointing“down
- Optic nerves are very atrophic.
- Schizencephaly (50% of the cases).
- Corpus callosum agenesis may also be seen.
- +/- Thin pituitary stalk
- Posterior pituitary ectopia



E- Middle Interhemispheric (MIH) Variant

- Sylvian fissures connect across midline over vertex (86%)
- Interhemispheric fusion of posterior frontal/parietal lobes
- Normal separation of anterior frontal/occipital lobes
- Non-cleavage of thalami > basal ganglia
- Heterotopias and cortical dysplasias common (86%)



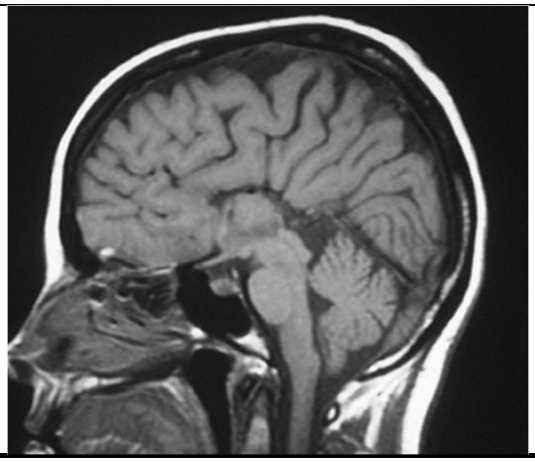


2) Dysgenesis of Corpus Callosum

- Growth anterior to posterior starting at the genu.
- Myelination from posterior to anterior.

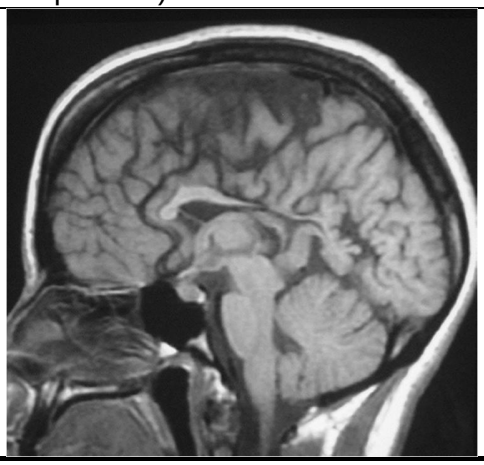
Complete Agenesis:

Corpus callosum is totally absent.

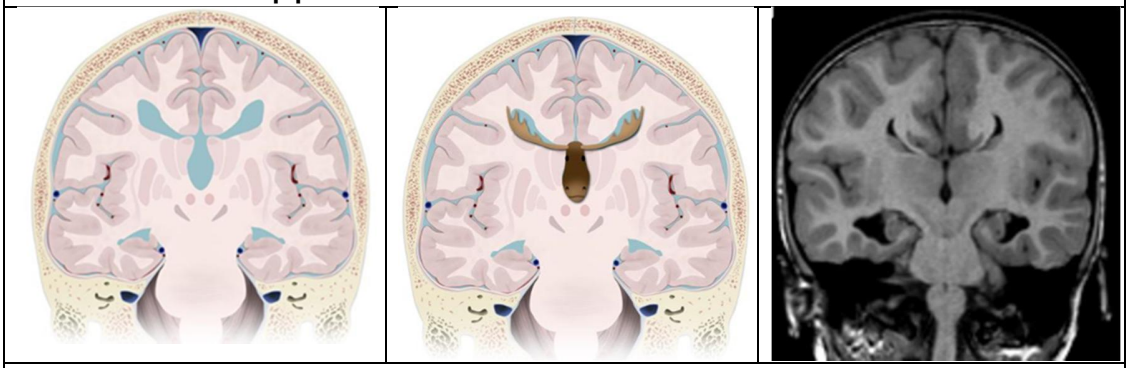


Partial Agenesis (hypoplasia):

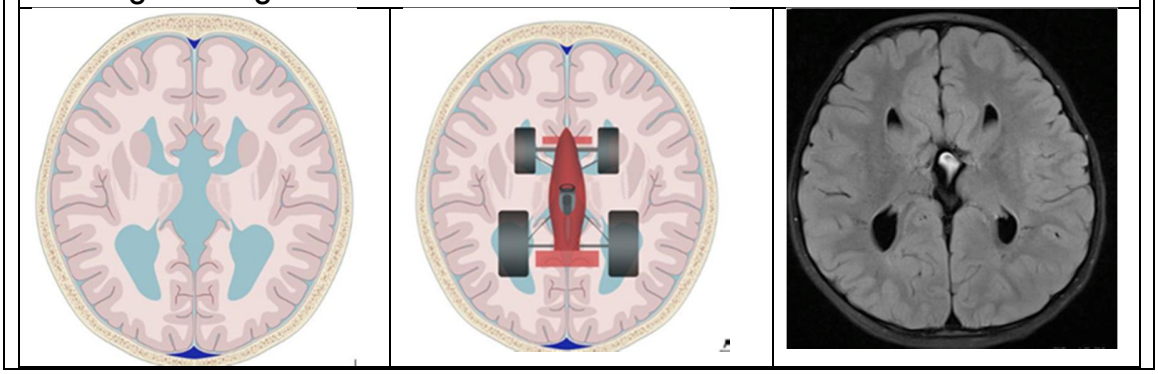
- Anterior portion (Genu + anterior body) is formed
- Posterior portion (Posterior body and splenium) + Rostrum is not formed.

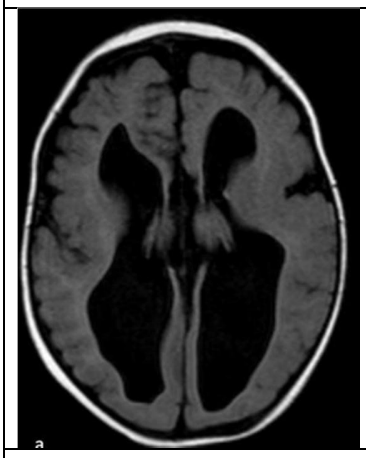
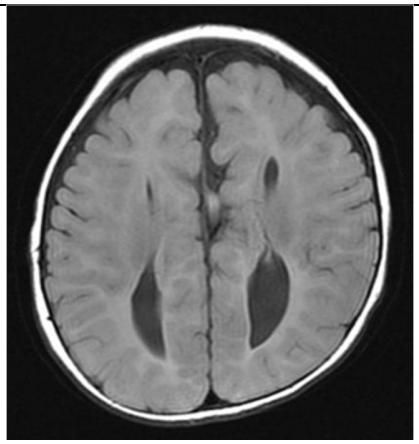
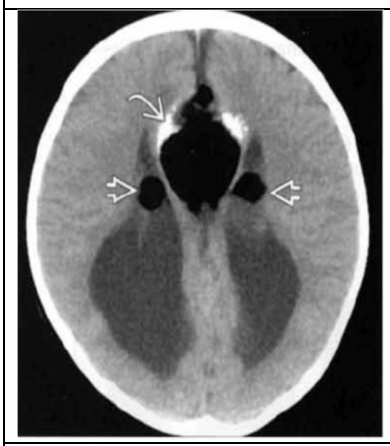
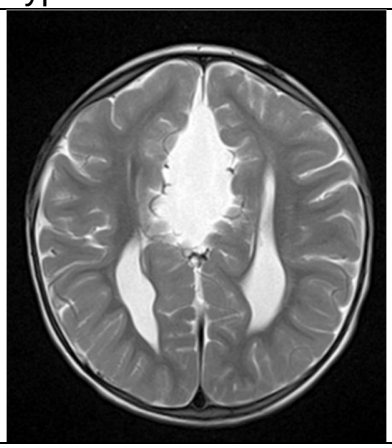


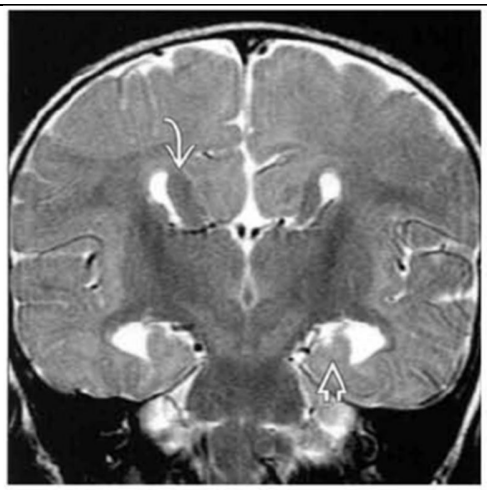
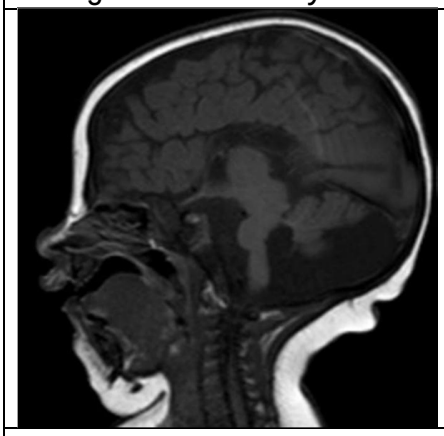
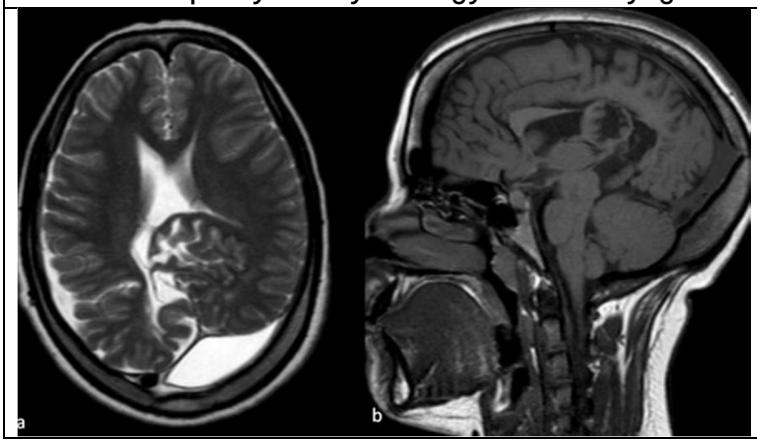
Moose Head Appearance



Racing Car Sign



<table>
<tr><td colspan="2">Axial => Lateral Ventricles:
Parallel (non-converging) - Widely separated
Occipital horns often dilated (Colpocephaly)
Pointed frontal horns</td></tr>
<tr><td>  </td><td>  </td></tr>
<tr><td colspan="2">Midline:</td></tr>
<tr><td>Calcified lipoma</td><td>High-riding 3rd ventricle
Interhemispheric Cyst:
Type 1: Not communicate with ventricle
Type 2: Communicate</td></tr>
<tr><td>  </td><td>  </td></tr>
<tr><td>Sagittal
Absent CC
Absent cingulate gyrus
Radially arrayed gyri "point to" 3rd ventricle</td><td>Coronal:
Trident-shaped anterior horns
Elongated foramina of Monro
Keyhole temporal horns
Vertical hippocampi
Probst bundles: Represent non-crossing commissural fibers</td></tr>
</table>

	longitudinally oriented WM tracts, indent medial ventricular walls
	
CC Agenesis + Dandy-Walker	
	
Morsier Syndrome =	
Septooptic dysplasia = Total absence of septum pellucidum + Schizencephaly + Polymicrogyria + CC Dysgenesis	
	

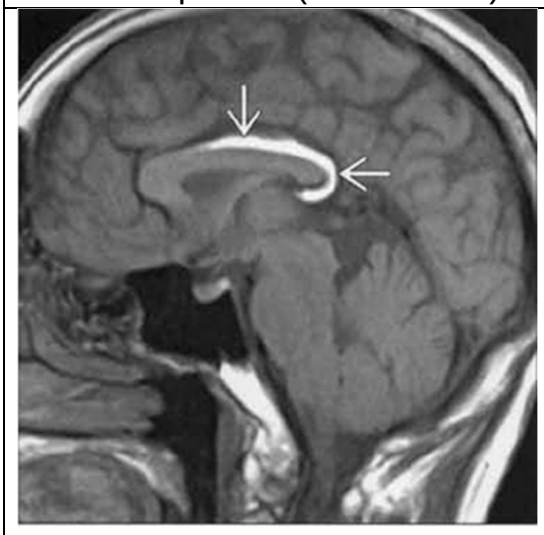
Intracranial Lipoma = Lipomatous Hamartoma

Locations:

- Deep interhemisphereic (Callosal)
- Suprasellar
- Qudrigminal
- CPA
- Sylvian

Radiological Features:

Well defined extra-axial lesion
CT: Low density lesion
T1: High signal
T2: High signal
STIR: loss of signal
T1 FS: Loss of signal
T1 +C FS: No enhancement

Interhemisphereic (Peri-Callosal)	Qudrigminal Lipoma
	

Stage III: Neuronal Migration and Proliferation Anomalies

Disorders of cortical formation according to stages

Stage	Cause	Disorder
Proliferative	Decreased proliferation	Microlissencephaly
	Increased proliferation	Hemimegalencephaly
	Abnormal proliferation	Focal cortical dysplasia
Migration	Undermigration	Complete (classic) lissencephaly
	Overmigration	Congenital muscular dystrophy
	Ectopic migration	Heterotopia
Organization	Deranged organization	Polymicrogyria
		Schizencephaly

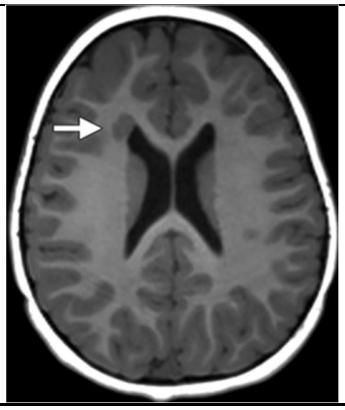
1) Gray Matter Heterotopia

Types:

- ❖ Subependymal heterotopia (most common)
- ❖ Band heterotopia ("double cortex")
- ❖ Focal subcortical hetrotopia.

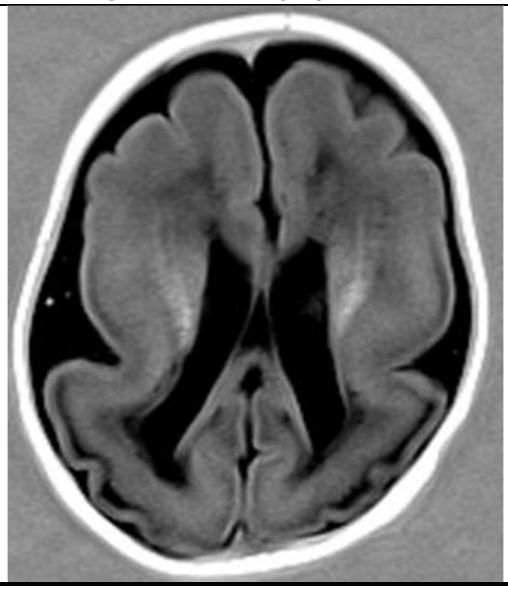
Radiological features:

- Linear or nodular or band like
- Subcortical or subependymal
- MRI: Isointense to gray matter with all pulse sequences

Subependymal GM Heterotopia	Band GM Heterotopia	Focal GM heterotopia
		

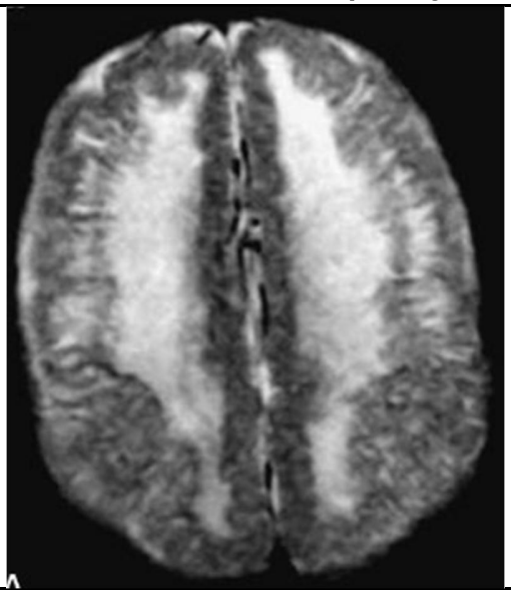
2) Lissencephaly Type I (Classic) Subcortical Band Heterotopia Spectrum

- Smooth cerebral surface
- Cortex is markedly thickened
- Few shallow sulci
- Vertically oriented shallow Sylvian fissures,
- Hour glass or figure-8 appearance of the brain on axial imaging
- Subcortical band heterotopia.
- CC agenesis or dysgenesis



3) Lissencephaly Type II = Cobblestone Lissencephaly

- Multi-nodular surface to the cortex (cobblestone)
- Lack of normal sulcation
- Small Sylvian fissure
- Hour glass or figure-8 appearance of the brain on axial imaging



4) Polymicrogyria

Neurons reach the cortex but are abnormally distributed into multiple small gyri like dimples on the surface of a basketball.
Clinically Seizures, developmental delays.

Location:

Predilection for the perisylvian region which is involved in 80% of patients and bilateral involvement is common (60%).

MRI

Gyri: multiple small, narrow, crowded gyri, has signal characteristics similar to normal grey matter.
GM/WM junction is always irregular.
Sulci are shallow.
Associated with schizencephaly.

5) Focal Transmantle Cortical Dysplasia

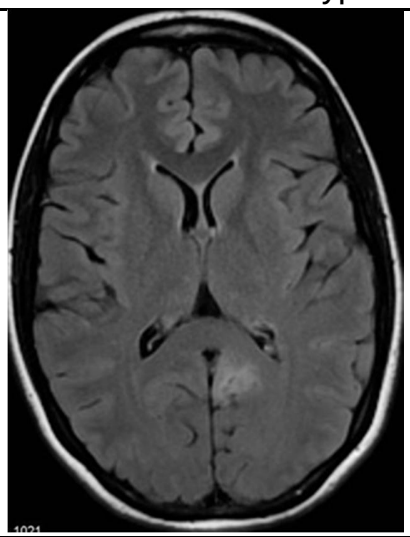
- Presence of abnormal neurons and glial cells within a localized region of the cerebral cortex.
- FCD is one of most common causes of intractable extra-temporal epilepsy in children.

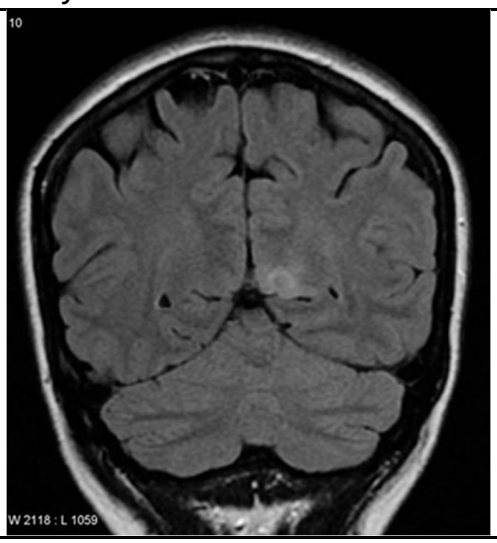
2 Types:

Type I: Non-Taylor
Type II: Taylor dysplasia or focal cortical dysplasia of Taylor (FCDT)

MRI findings:

- Cortical + Subcortical T2 hyperintensity,
- Focal cortical thickening.
- Blurring of GM/WM junction.
- MRI/FDG PET: hypometabolic activity

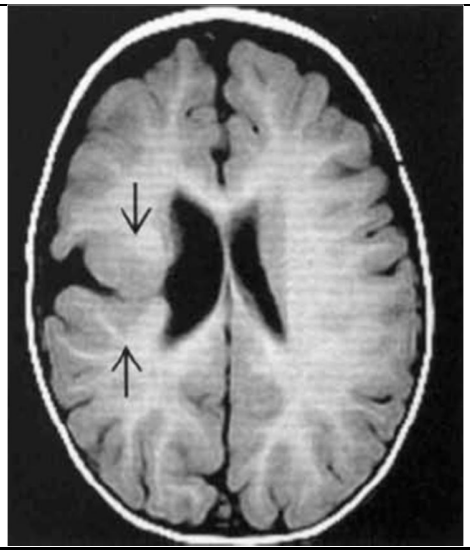
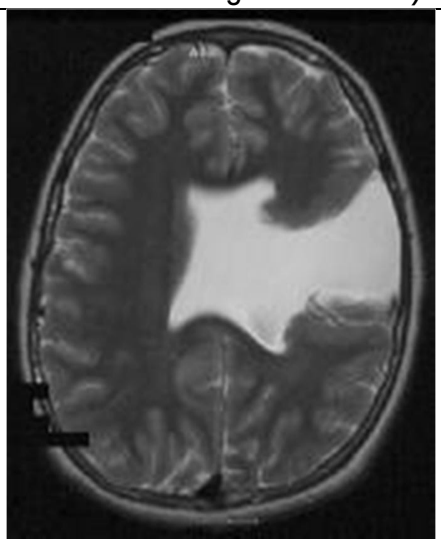




6) Schizencephaly

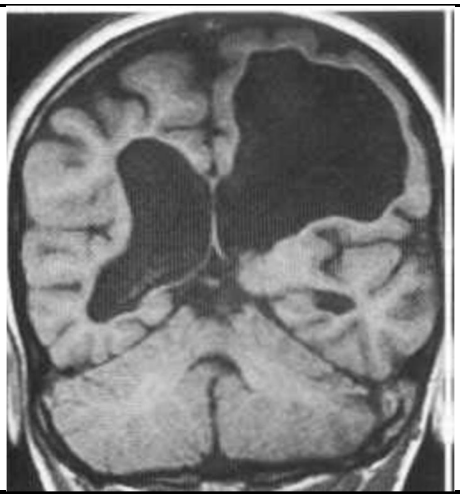
Clefts extending from pial surface of cerebral mantle to ventricle lined by polymicrogyric cortex

Two Types:

Closed lip (mild, no CSF within)	Open lip (contains CSF, severe with cortical defects and large ventricles).
	

D.D: PORENCEPHALY

- Intra-axial well defined cyst
- Communicate with lateral ventricle
- Lined by grey matter



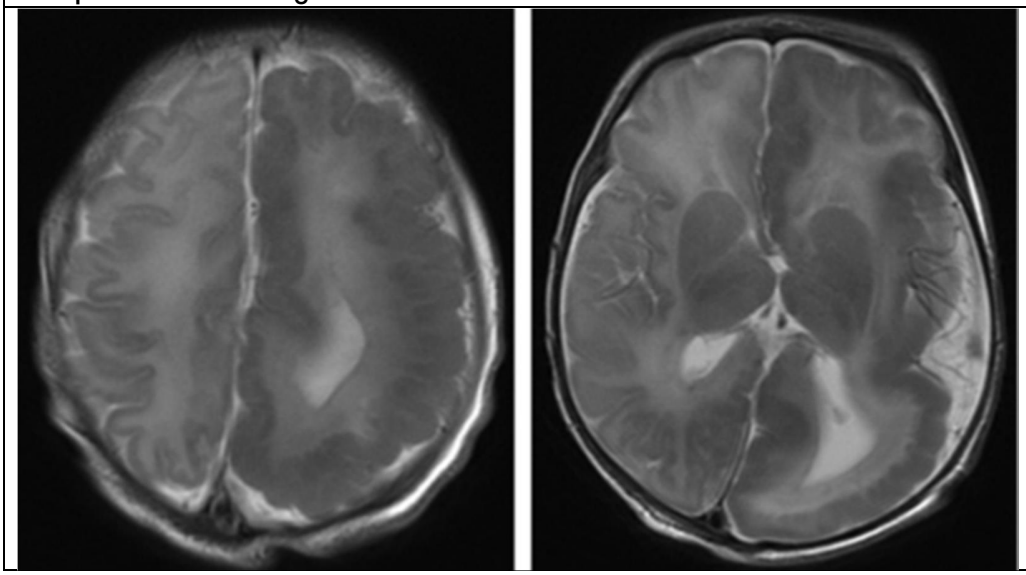
7) Hemimegalencephaly

Hemimegalencephaly represents partial or total hamartomatous overgrowth of a cerebral hemisphere.

- Isolated entity or
- Association with other syndromes:
  - Neurofibromatosis type 1
  - Tuberous sclerosis
  - Epidermal nevus syndrome
  - Hypomelanosis of Ito
  - Klippel-Trénaunay- Weber syndrome.

Imaging findings:

- Unilateral cortical thickening involving all or part of the cerebral hemisphere.
- Broad featureless gyri and shallow sulci
- Ipsilateral white matter changes secondary to hypermyelination.
- Ipsilateral enlargement of the lateral ventricle.



Congenital Abnormalities of Posterior Fossa

I) Predominantly Cerebellar Involvement:

A) Predominantly Vermian:

- Posterior fossa Cystic Lesions:
  - Dandy-Walker Malformation
  - Blake pouch cyst
  - Mega cisterna magna
  - Arachnoid cysts
  - Isolated vermian hypoplasia
- Rhombencephalosynapsis

B) Global Cerebellar:

- Malformations of cortical development:
  - Lissencephaly, Polymicrogyria, periventricular nodular heterotopia (FLNA), and primary microcephaly
- Macrocerebellum
- Cerebellar dysplasia:
  - Chudley-McCullough syndrome
- Global cerebellar hypoplasia
  - Multiple genetic syndromes (eg, Ritschcer-Schinzel syndrome, CHARGE syndrome, Delleman syndrome)
- Congenital CMV infection

C) Unilateral Cerebellar:

- Isolated unilateral cerebellar hypoplasia, PHACES syndrome, COL4A1 mutations, cerebellar cleft

II) Cerebellar & Brain Stem Involvement:

- Pontocerebellar hypoplasia (PCH)
- Glycosylation type 1a congenital disorder
- CASK mutations
- Joubert syndrome
- Cerebellar agenesis
- Congenital muscular dystrophies (CMD)
- Vanishing cerebellum in Chiari 2 malformation
- Cerebellar disruption secondary to prematurity

III) Predominantly brainstem involvement

- Pontine tegmental cap dysplasia
- Horizontal gaze palsy and progressive scoliosis

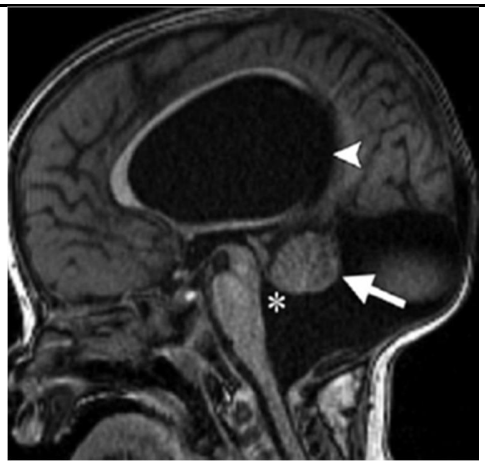
IV) Predominantly midbrain involvement

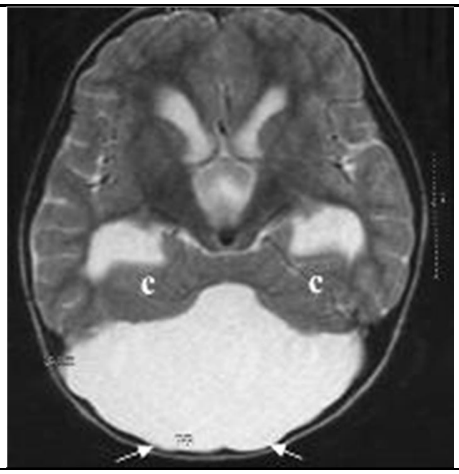
- Dysplasia of the diencephalic-mesencephalic junction

❖ Posterior Fossa Cystic Lesions:

1) Dandy Walker Malformation:

- Enlarged posterior fossa.
- Cystic dilatation of 4th Ventricle
- Wide communication between cyst posteriorly and 4th ventricle => "keyhole" appearance.
- Hypoplasia of the vermis + superiorly rotated vermian remnant
- Cerebellar hemispheres are typically displaced anterolaterally.
- The tentorium, torcular and transverse sinuses are elevated



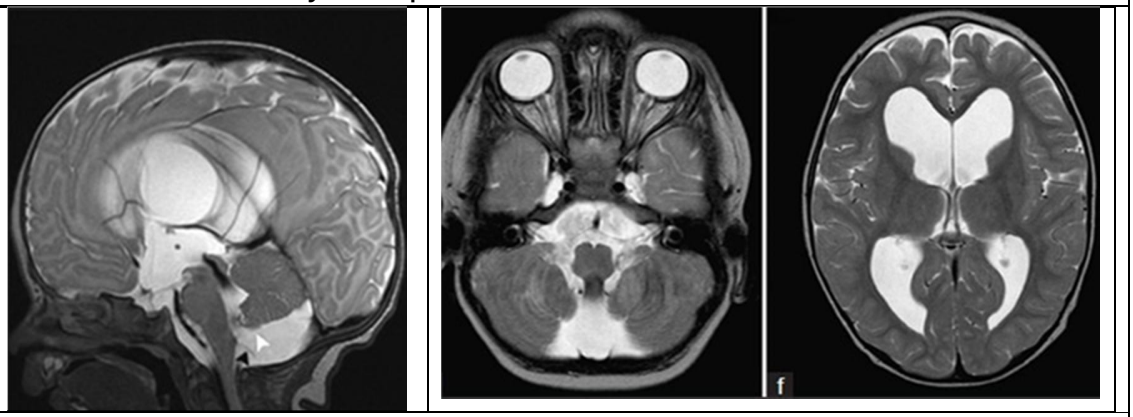


2) Mega Cisterna Magna (MCM)

- Normal cerebellar hemispheres and vermis
- Large retrocerebellar CSF collection communicates with 4th ventricle
- Dural fold (arrow)
- Normal 4th ventricle
- Occasionally, quite enlarged posterior fossa

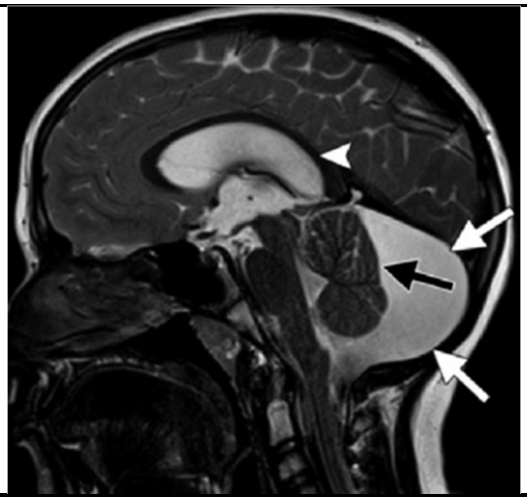
3) Blake Pouch Cyst (BPC)

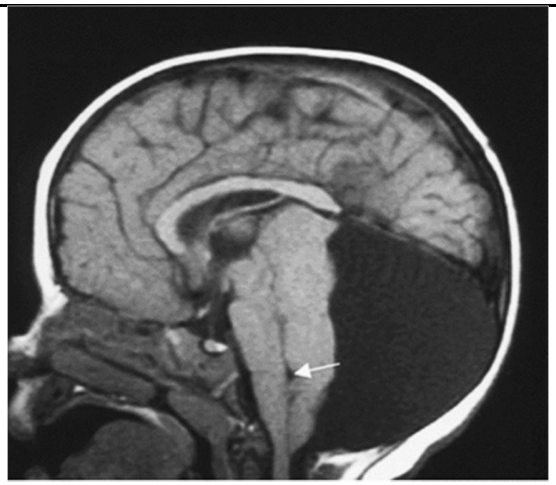
- Cystic appearing structure that represents posterior ballooning of the inferior medullary velum into the cisterna magna
- Infravermian cyst = Below and posterior to vermis
- Communicates with open fourth ventricle.
- Don't communicate with cisterna magna.
- No vermian hypoplasia or rotation
- Cystic dilatation of 4th ventricle
- 4th ventricular choroid plexus continuing in the roof of the cyst
- Tetraventricular Hydrocephalus



4) Posterior Fossa Arachnoid Cyst (PFAC)

- Location:
    - Retrocerebellar = inferior or posterior to the vermis
    - Supravermian = cranial to the vermis in the tentorial hiatus
    - CPA cistern = anterior or lateral to the cerebellar hemispheres
    - Prepontine cistern = anterior to the brainstem
- Well defined extra-axial CSF collection / cyst
- The cyst walls are usually too thin to be visualized at MRI
- Not communicate with the fourth ventricle
- Not communicate with the subarachnoid space
- Mass effect on the dorsal aspect of a normal-appearing vermis and cerebellum
- Normal but displaced fourth ventricle
- Usually enlarged posterior fossa
- Scalloping of occipital bone
- +/- Supratentorial Hydrocephalus





5) Isolated Vermian Hypoplasia (IVH)

- Partial absence of the inferior portion of the cerebellar vermis on midsagittal image.
- Normal size and architecture of:
  - Remainder of the vermis
  - Cerebellar hemispheres
  - 4th ventricle
  - Posterior fossa

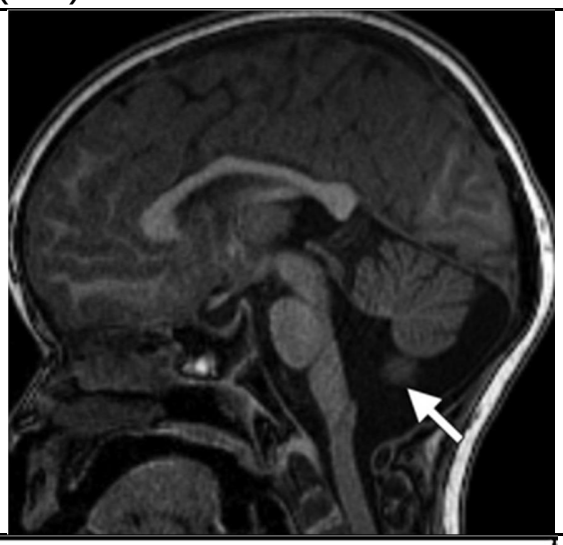
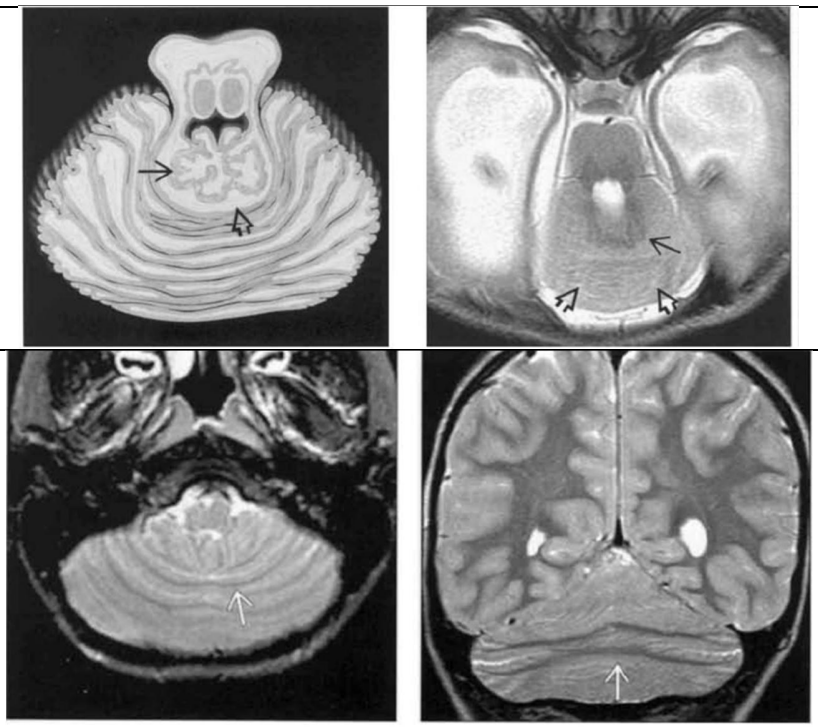


Table 3: Key Neuroimaging Features of DWM versus Other Cystic Posterior Fossa Malformations

Malformation	Vermis	Fourth Ventricle	Posterior Fossa	Hydrocephalus	Occipital Bone Scalloping*
DWM	Hypoplastic	Enlarged	Enlarged	Yes (in most patients)	No
IVH	Hypoplastic (inferior portion)	Enlarged	Normal	No	No
BPC	Normal	Enlarged	Normal	Yes	No
MCM	Normal	Normal	Inconsistently enlarged	No	Possible
PFAC	Normal	Normal or reduced	Normal	Possible	Yes

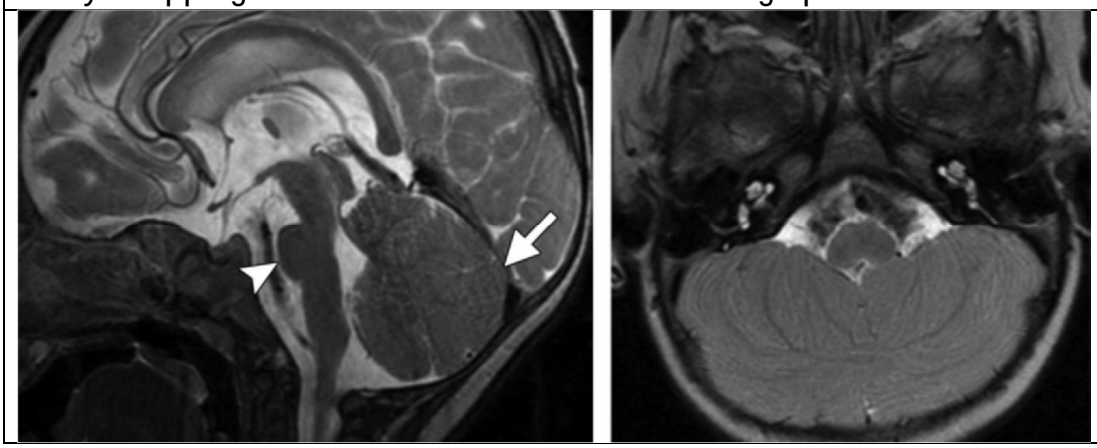
❖ Rhombencephalosynapsis

- No vermis + Cerebellar hemispheres are fused
- Congenital fusion of cerebellar hemispheres, dentate nuclei, and superior cerebellar peduncles
- Abnormal transverse orientation of the cerebellar foliae



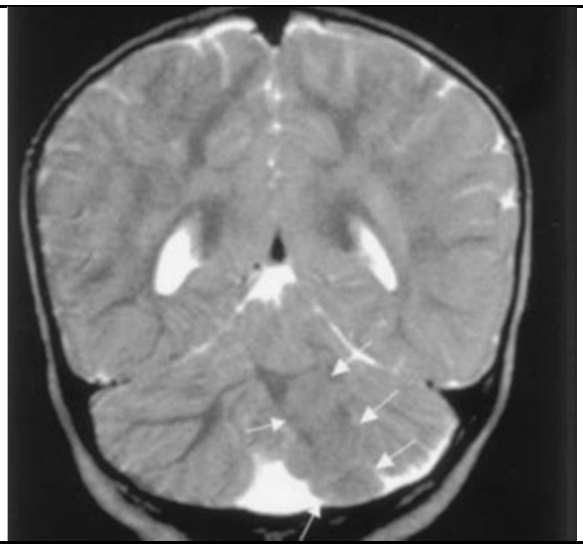
❖ Macrocerebellum

- An extremely rare condition
- Large cerebellum with preservation of its architecture and shape.
- The cerebellar hemispheres may expand into adjacent anatomic regions by wrapping around the brainstem or herniating upward or downward.



❖ Cerebellar Dysplasia

- Abnormal foliation
- Abnormal fissuration (including defective, enlarged, or vertical fissures),
- Abnormal white matter arborization
- Loss of GM/WM junction.
- Supratentorial findings:
    - o Migrational abnormalities
    - o Callosal dysgenesis
    - o Clastic lesions



❖ Chudley-McCullough Syndrome

- Cerebellar dysplasia in the inferior cerebellar hemispheres
- Severe sensorineural hearing loss
- Mild developmental delay
- Partial agenesis of the corpus callosum
- Periventricular heterotopia

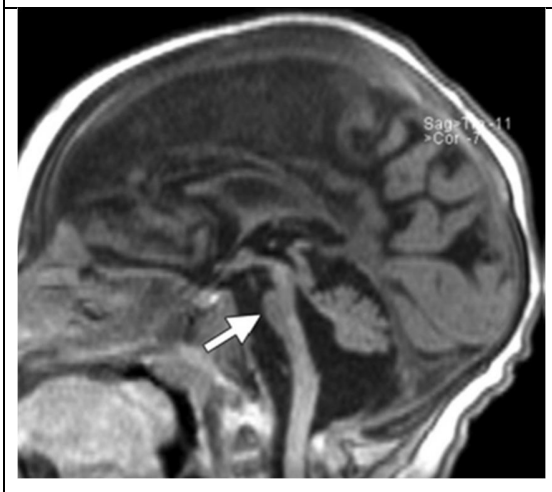
❖ Cerebellar and Brainstem Malformations

1) Pontocerebellar Hypoplasia (PCH)

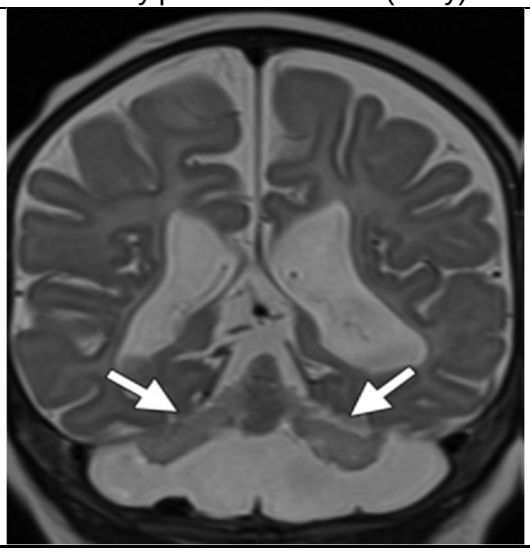
Table 4: Subtypes of PCH

Subtype	Main Features	Genotype
1	Involvement of the anterior horn cells	*VRK1*, *TSEN54*, *RARS2*, *EXOSC3*
2	Chorea/dystonia; rarely, spasticity only	*TSEN54* (vast majority of cases); *TSEN2*, *TSEN34*
3	Optic atrophy, nonprogressive course	Locus on 7q11–21
4	Severe neonatal course with hypertonia and hypoventilation, absent inferior olivary prominence at autopsy	*TSEN54*
5	Lethal course with neonatal death, more pronounced involvement of the vermis than of the hemispheres	*TSEN54*
6	Lactic acidemia, high lactate level in CSF	*RARS2*
7	Micropenis with no palpable gonads	Unknown
8	Proportionate involvement of the vermis and hemispheres, nonprogressive course	*CHMP1A*
9	Dysgenesis of the corpus callosum	*AMPD2*
10	Dysmorphic features and axonal sensorimotor neuropathy (inconsistent)	*CLP1*

Hypoplasia of the pons (arrow) and cerebellar vermis



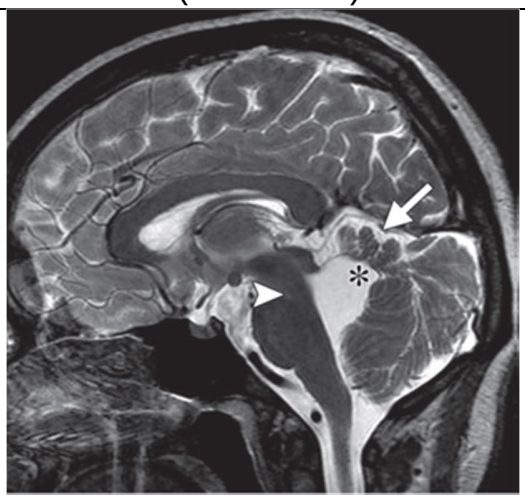
“Dragonfly” appearance =
Flattened cerebellar hemispheres (wings) + Relatively preserved vermis (body)



2) Joubert's Syndrome

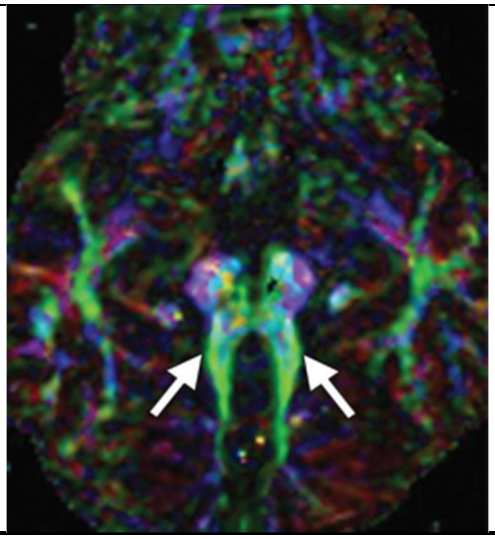
- Molar tooth sign.
- Elongated, thickened, and horizontally oriented superior cerebellar peduncles
- Deepened interpeduncular fossa

- Hypoplasia and dysplasia of the vermis (arrow)
- Enlargement of 4th ventricle.
- Narrow pontomesencephalic isthmus (arrowhead)



<u>Axial color coded FA map</u>

- Horizontal orientation of the superior cerebellar peduncles (green [arrows])
- Absence of decussation of the superior cerebellar peduncles.



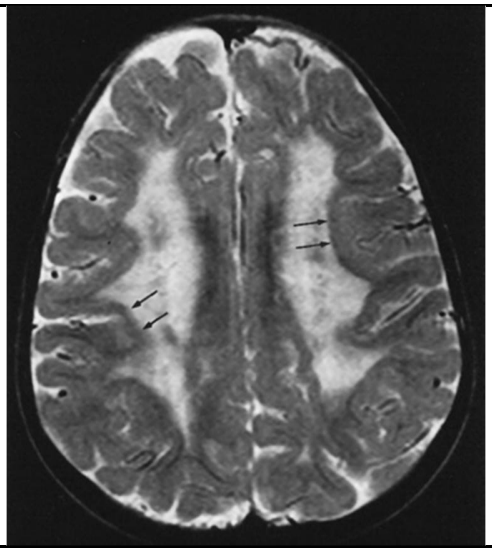
3) CONGENITAL MUSCULAR DYSTROPHY (CMD)

- CMD 1: "Pure" CMD
  - Merosin-positive CMD
  - Merosin-negative CMD
- CMD 2: Fukuyama (FCMD) = least severe
- CMD 3: Muscle-eye-brain (MEB)
- CMD 4: Walker-Warburg syndrome (WWS) = most severe

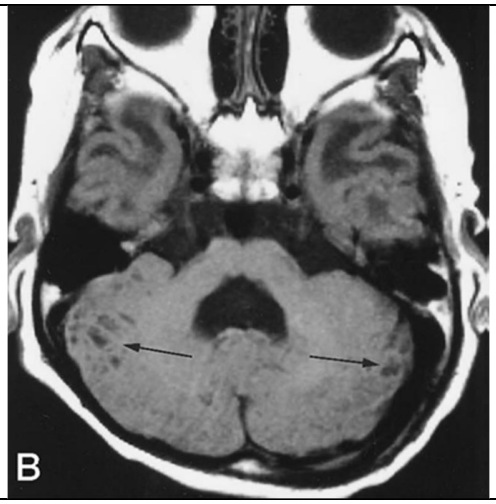
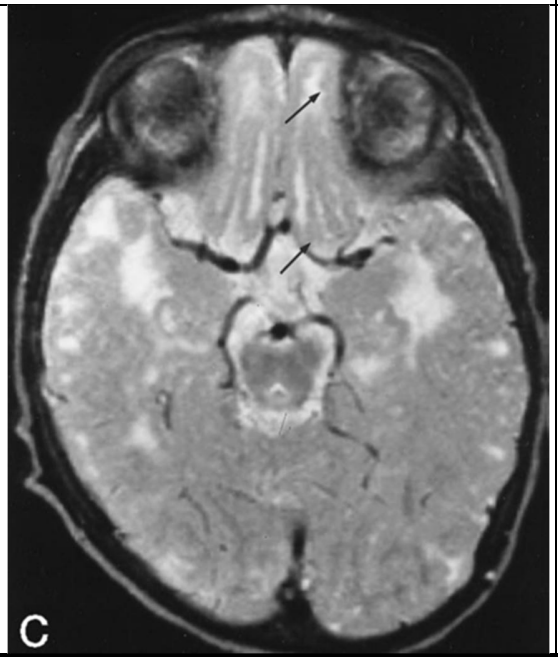
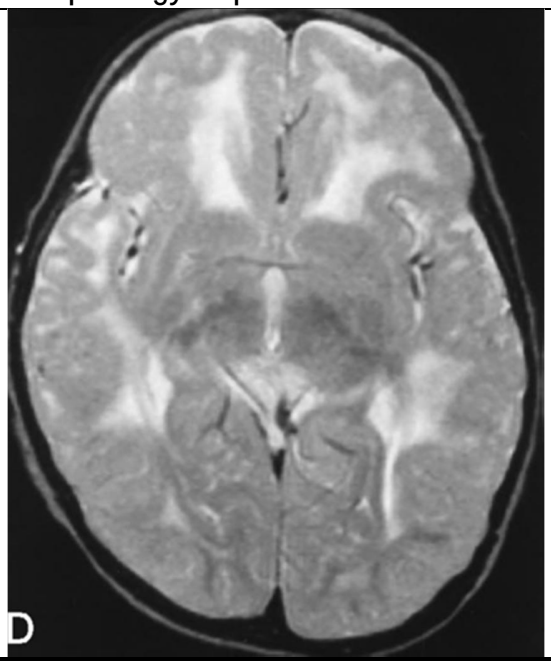
- The congenital muscular dystrophies (CMDs) are a heterogeneous group of disorders characterized by hypotonia, weakness, and, frequently, congenital contractures in conjunction with dystrophic changes of muscle as established by muscle biopsy

a) Pure CMD

- T2 hyperintense of the cerebral white matter, interpreted as hypomyelination
- Mild hypogenesis of the cerebellar vermis
- Slightly small pons.
- The cerebral and cerebellar cortices and the white matter of the brain stem and cerebellum appeared normal.



b) Fukuyama CMD

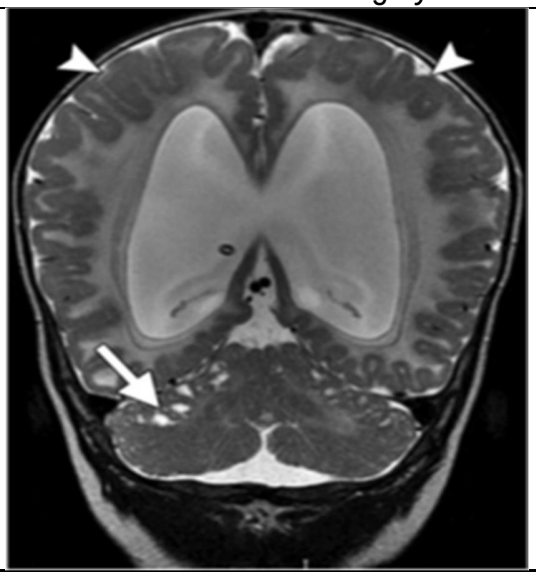
• Marked pontine hypoplasia (straight arrow) • Fusion of the midbrain colliculi (curved arrow) • Hypoplasia of inferior cerebellar vermis	• Enlarged 4th ventricle • Vermian hypoplasia • Cystic type of cerebellar polymicrogyria • The cysts (arrows) primarily in the lateral cerebellar cortex
	
Inferomedial frontal cortex irregular cortical-white matter junction, suggesting polymicrogyria.	T2 hyperintense of the central cerebral white matter Normal appearing occipital and temporal gyral patterns.
	

c) Muscle-Eye-Brain MEB

Early clinical symptoms include muscular hypotonia and poor visual contact.

- Hypoplasia of the vermis
- Flattening of the ventral pons
- Dysmorphic tectum and midbrain (arrow)
- Abnormal concave posterior border of the brainstem (*)
- Enlarged 4th ventricle, and supratentorial ventriculomegaly (arrowhead).

- Cerebellar hypoplasia
- Multiple bilateral subcortical cysts in the cerebellar hemispheres (arrow), cerebellar dysplasia.
- Generalized polymicrogyria (arrowheads)
- Abnormal signal intensity of the supratentorial white matter
- Absence of the septum pellucidum,
- Marked ventriculomegaly.



d) Walker-Warburg Syndrome

HARDE syndrome

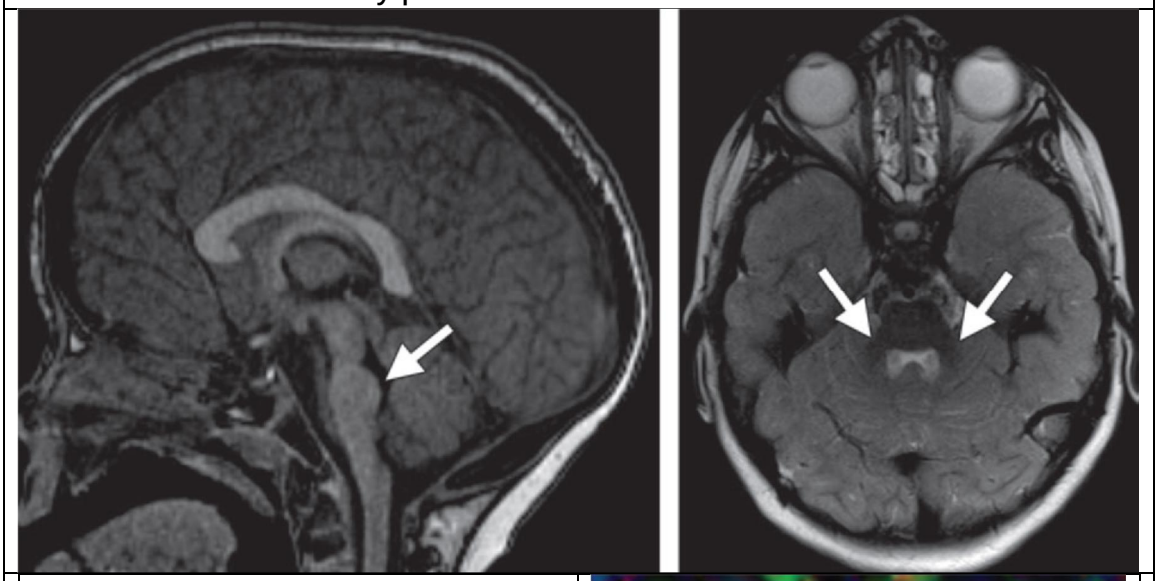
- Hydrocephalus : almost always present
- Agyria (cobblestone lissencephaly / lissencephaly type II)
- Retinal dysplasia
- Dandy Walker classic / variant
- Encephalocele

• Z-shaped" hypoplastic brainstem. • Dorsal pontomesencephalic Kink • The belly of the pons is notched • Cerebellum is severely hypoplastic and rotated.	• Cobblestone lissencephaly / lissencephaly type II • Hydrocephalus .

❖ Predominantly Brain Stem Malformations

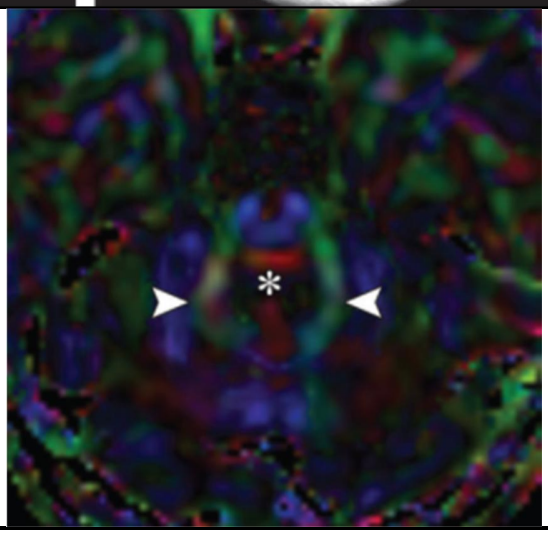
1) Pontine Tegmental Cap Dysplasia:

- Flattened ventral pons
- A cap covering the dorsal pons and protruding into 4th ventricle.
- Partial absence of the middle cerebellar peduncles, vermian hypoplasia,
- A molar tooth–like aspect of the pontomesencephalic junction
- Absent inferior olivary prominence



<u>Axial color-coded FA map:</u>

- Absence of transverse pontine fibers in the ventral and middle pons (horizontal red band)
- An ectopic band of fibers (horizontal orientation) dorsal to the pons (*)
- Small middle cerebellar peduncles (anteroposterior orientation) (arrowheads).



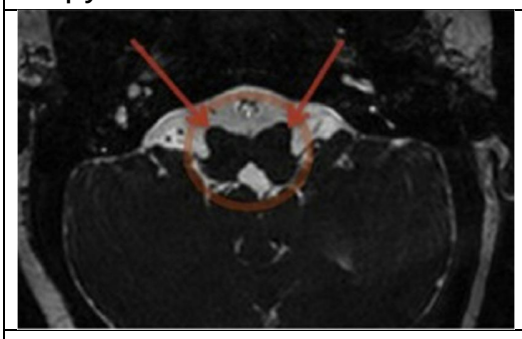
2) Horizontal Gaze Palsy with Progressive Scoliosis (HGPPS):

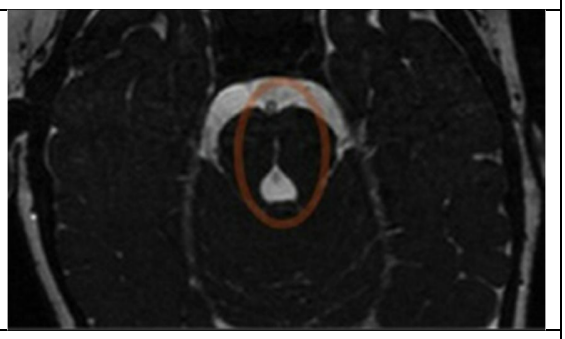
- DTI typically shows the absence of decussation of the corticospinal tracts, pontine sensory tracts, and superior cerebellar peduncles

<u>A butterfly-shaped medulla</u>

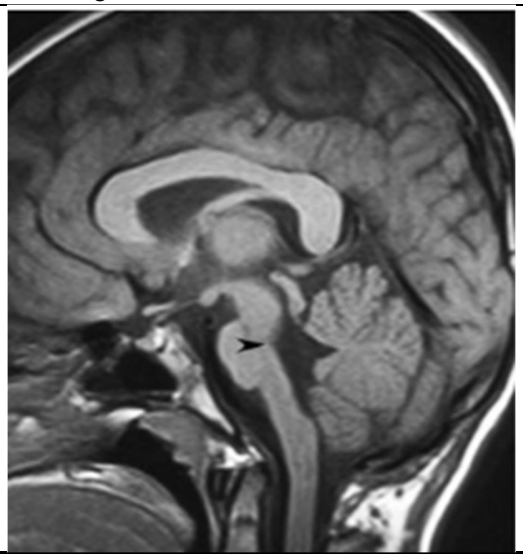
- Missing prominence of the gracile and cuneate nuclei.
- Prominent inferior olivary nuclei with respect to the medullary pyramids.

- The pons is hypoplastic
- Dorsal midline cleft
- Absence of the bulging contour of facial colliculi
- Floor of 4th ventricle tent-shaped





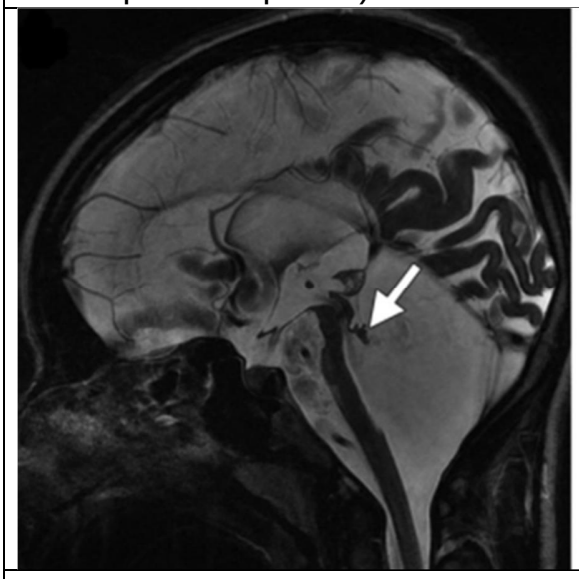
Hypoplastic pons + Medulla oblengata

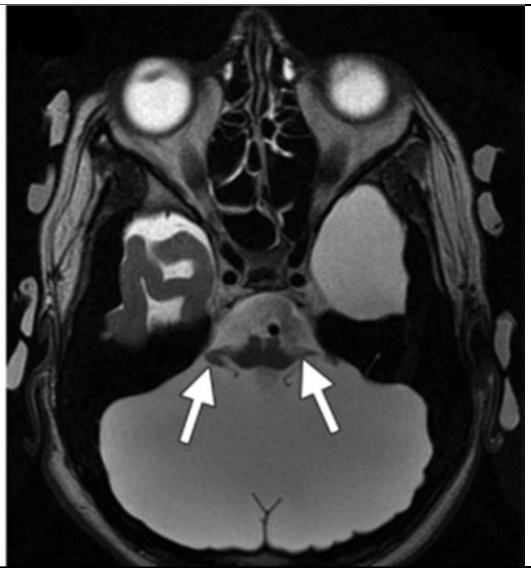


❖ Global Cerebellar

1) Cerebellar Agenesis

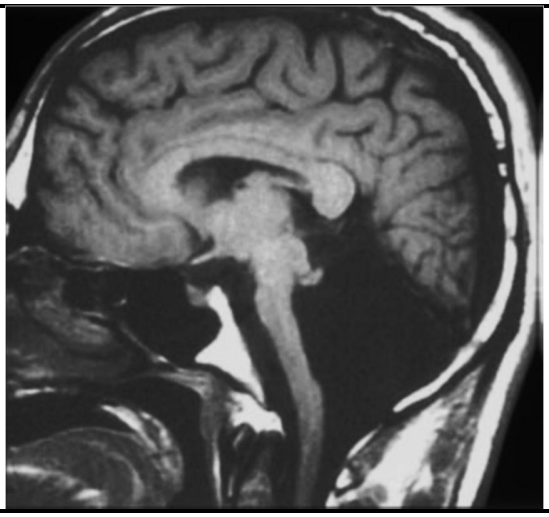
- Congenital malformation resulting from a genetically mediated pathomechanism (eg, mutations in PTF1A)
- Cerebellar disruption (eg, hemorrhage that occurs during gestation or in the perinatal period)





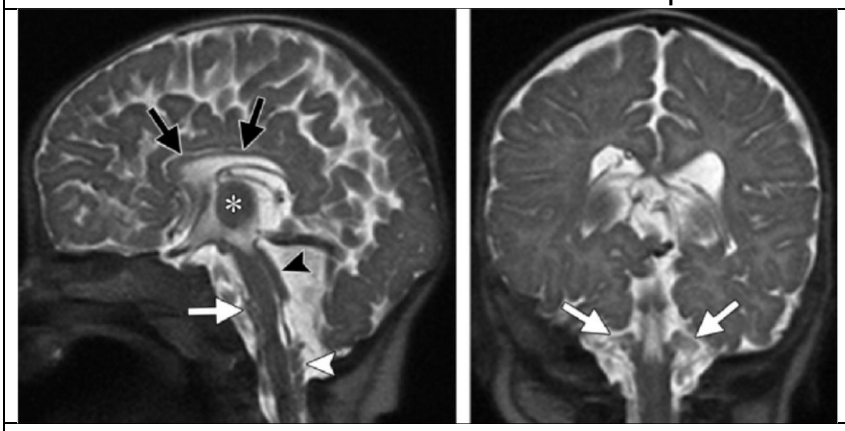
2) Global Cerebellar Hypoplasia

- Small cerebellum
- Normal-sized, posterior fossa.
- The pons and medulla are abnormally small.



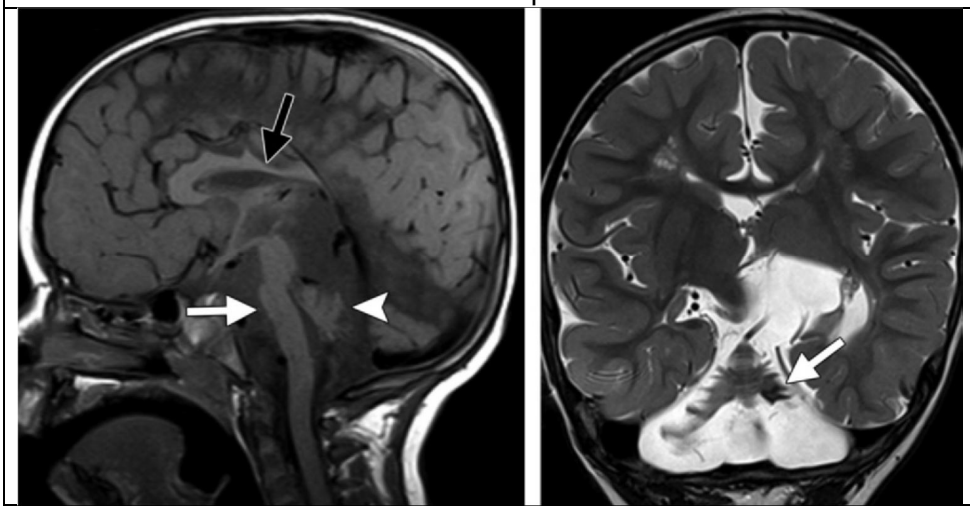
3) Vanishing Cerebellum in Myelomeningocele

Non–skin-covered myelomeningocele => Prenatal hindbrain herniation through the foramen magnum is believed to be secondary to a prolonged prenatal CSF leak => This herniation may result in parenchymal damage due to ischemia, presumably induced by mechanical distortion, and marked reduction in the size of the cerebellar hemispheres.



4) Cerebellar Disruption or Injury Secondary to Prematurity

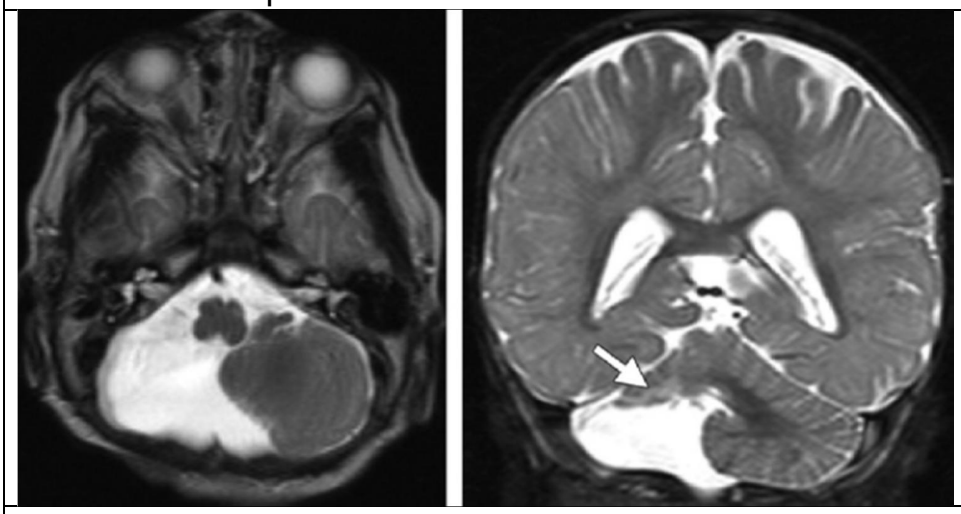
- Primarily destructive injuries include hemorrhage and ischemia and cause cerebellar tissue loss.
- Secondary lesions lead to underdevelopment of the cerebellum and are caused by:
    - Direct (eg, hemosiderin– blood products, glucocorticoid exposure, undernutrition)
    - Indirect (impaired transsynaptic trophic effects) effects on the cerebellum
- General perinatal risk factors for cerebellar injury in extremely premature neonates:
    - Prolonged ventilation, pressor support, posthemorrhagic hydrocephalus, and infratentorial hemosiderin deposition



❖ Unilateral Cerebellar:

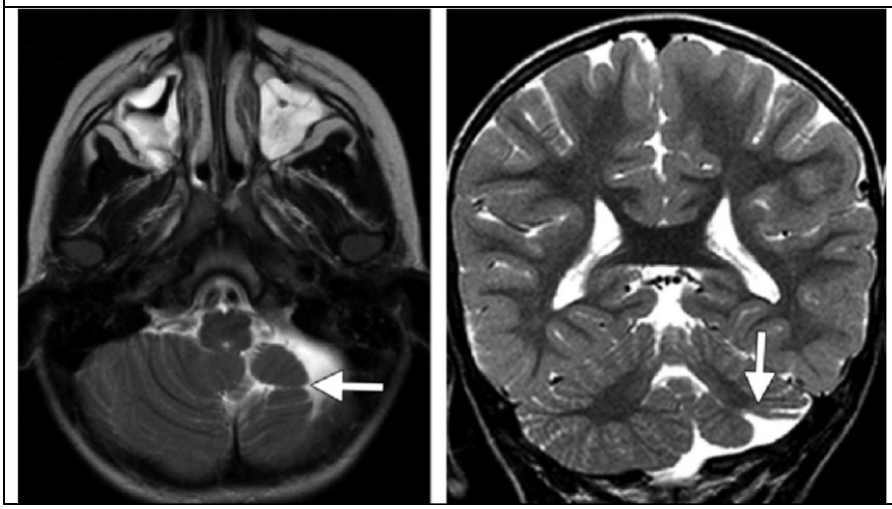
1) Unilateral Cerebellar Hypoplasia

- Volume loss in the cerebellar hemisphere and vermis
- Normal sized posterior fossa



2) Cerebellar Cle**ft**:

- A cleft extends from the surface of the hemisphere into the parenchyma, and may reach the fourth ventricle,
- Irregular gray matter–white matter junction
- Abnormal arborization of the white matter confined to the region adjacent to the cleft, along with volume reduction in the affected hemisphere
- But without involvement of the vermis



Normal Myelination

MRI is only imaging technique that assesses myelination

T1 WI: show myelination as increasing hyperintensity

- T1 is most sensitive sequence < 1 year of age

T2 WI: show myelination as increasing hypointensity

- T2 is most sensitive sequence between 1 and 2 y

FLAIR: Triphasic pattern

DWI: shows myelination as decreasing diffusibility.

FA map: show myelination as increasing FA

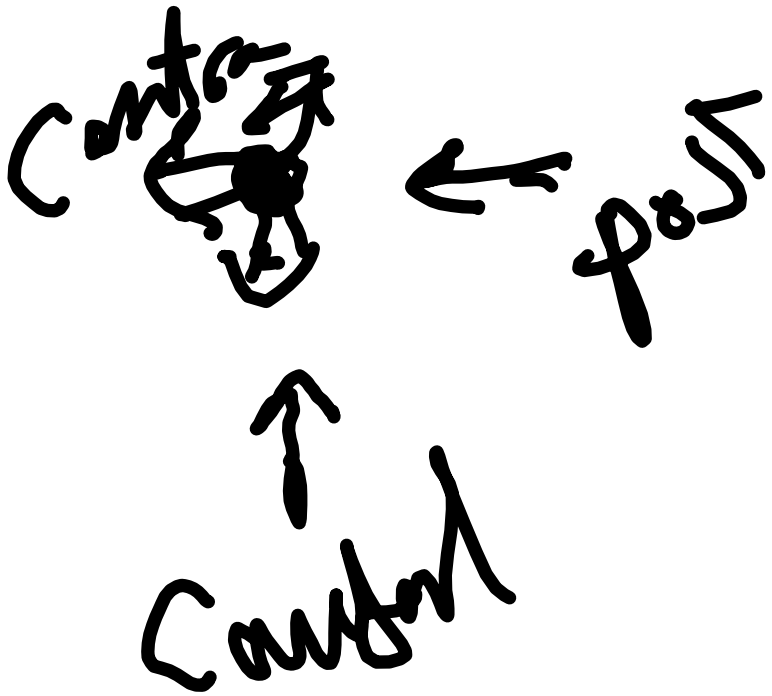
MTI: shows myelination as increasing MT

Normal myelination progress

→ From the caudal to the cranial aspect.

→ From the posterior to the anterior aspect.

→ From the center to the periphery.



Myelinated Structures at Birth

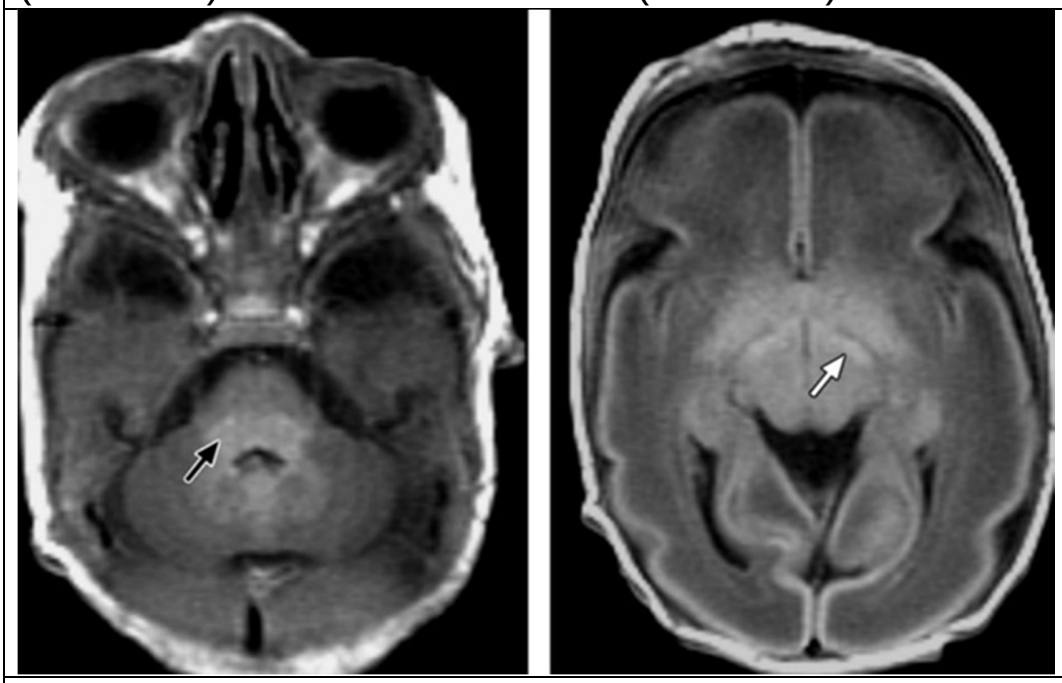
- dorsal brainstem
- ventrolateral thalamus
- lentiform nuclei
- central corticospinal tracts
- posterior limb of the internal capsule
- Middle cerebellar peduncle
- Optic nerve, chiasma and tract

Progression Of Myelination

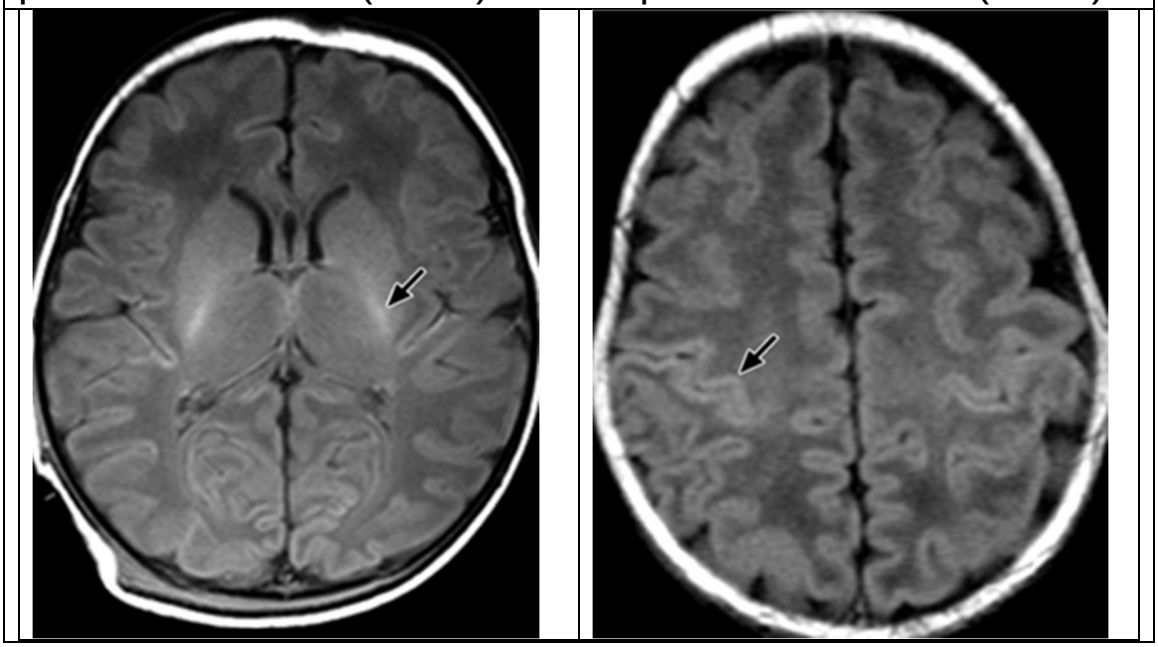
- The first change is increase in T1 signal, and later decrease in T2.
- **2-3 months:** anterior limb of IC becomes **T1** bright
- **3 months:** cerebellar WM tracts becomes **T1** bright
- **3-6 months:** splenium of corpus callosum becomes **T2** dark
- **6 months:** genu of corpus callosum becomes **T1** bright
- **8 months:** subcortical white matter becomes **T1** bright
- **8 months:** genu of corpus callosum becomes **T2** dark

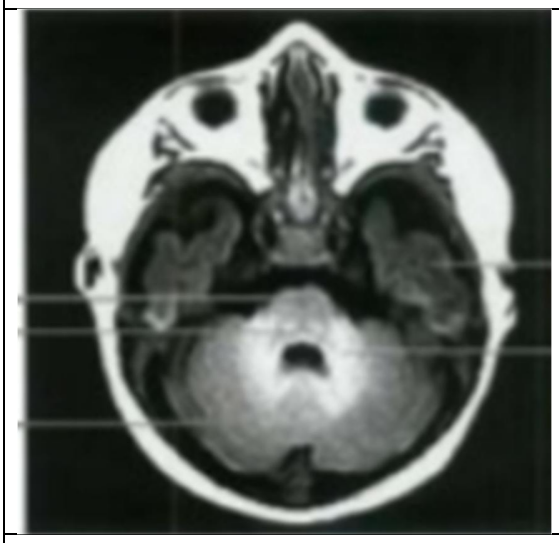
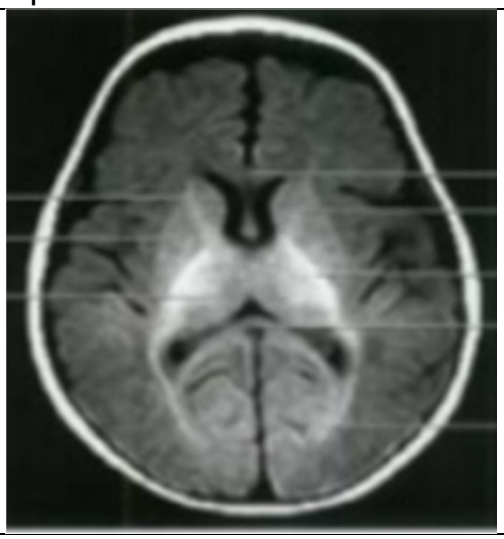
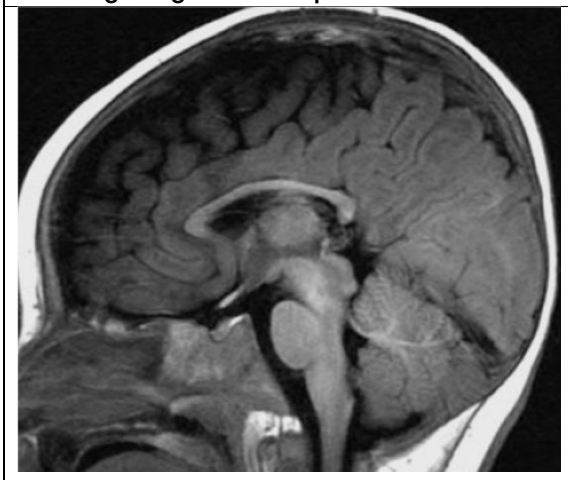
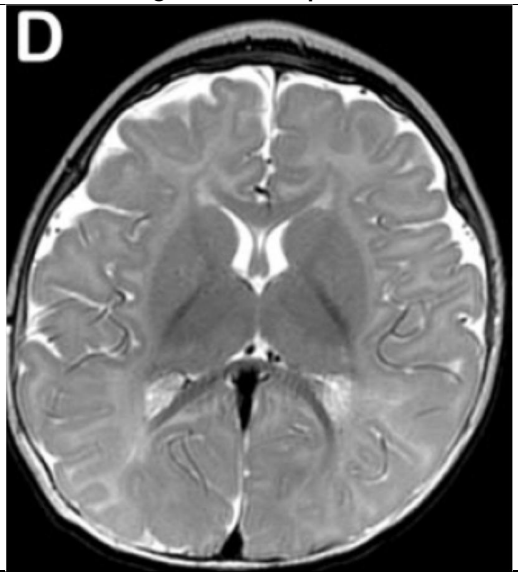
- **11 months:** anterior limb of internal capsule becomes **T2** dark
- **1 year 2 months:** occipital white matter becomes **T2** dark
- **1 year 4 months:** frontal white matter becomes **T2** dark
- **1 1/2 years:** majority of white matter becomes **T2** dark (except terminal myelination zones adjacent to frontal horns and periatrial regions)
- **2 years:** almost all of white matter becomes **T2** dark

T1: 28 w of gestation show myelination in the dorsal brainstem (arrow in a) and subthalamic nuclei (arrow in b).

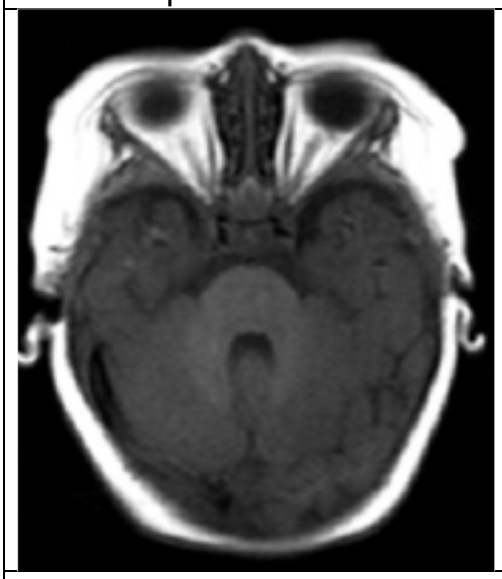
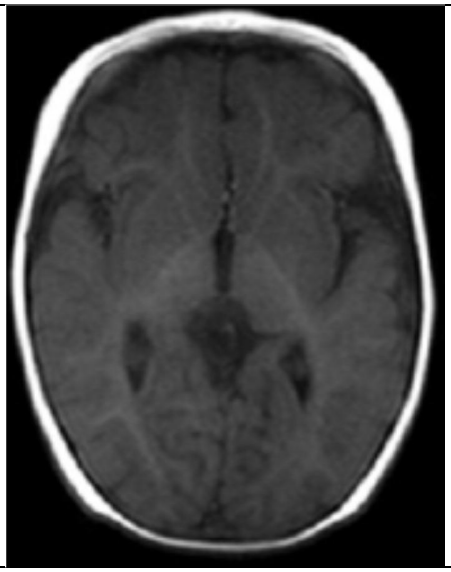


T1: 38 w of gestation show areas of hyperintense signal in the posterior limb of IC (arrow) and the perirolandic cortex (arrow).



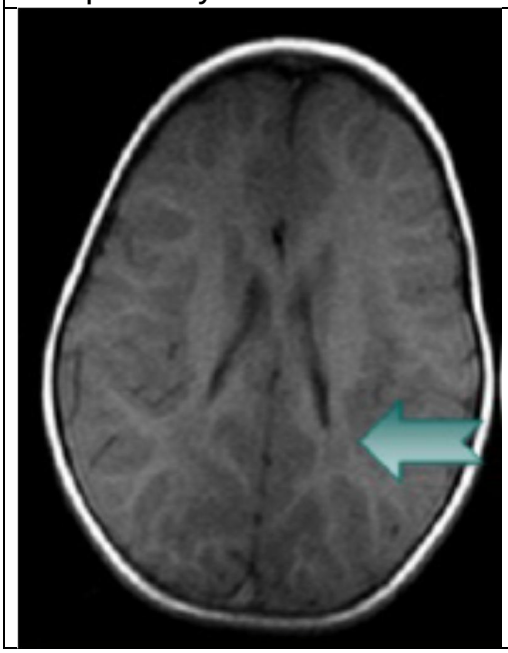
3m	
Middle cerebellar peduncles Cerebellar deep WM Ventral brain stem	Anterior limb of IC Corticospinal tract Optic radiation
	
6m	
T1: High signal of coprus callosum	T2: Low signal of corpus callosum
	

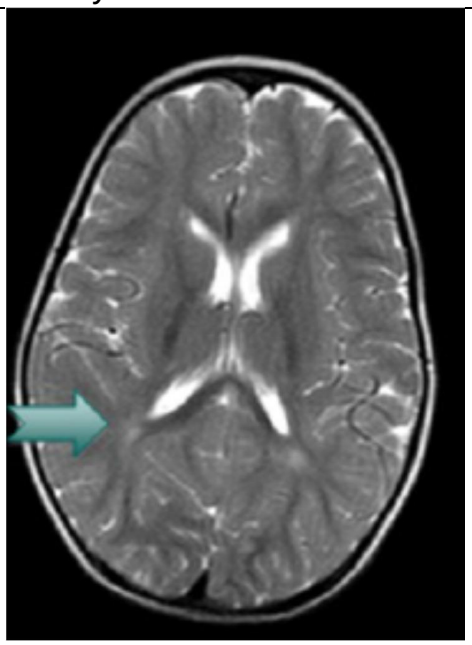
8-12m

Complete myelination of posterior fossa and posterior brain	Immature frontal WM
	

15-18m

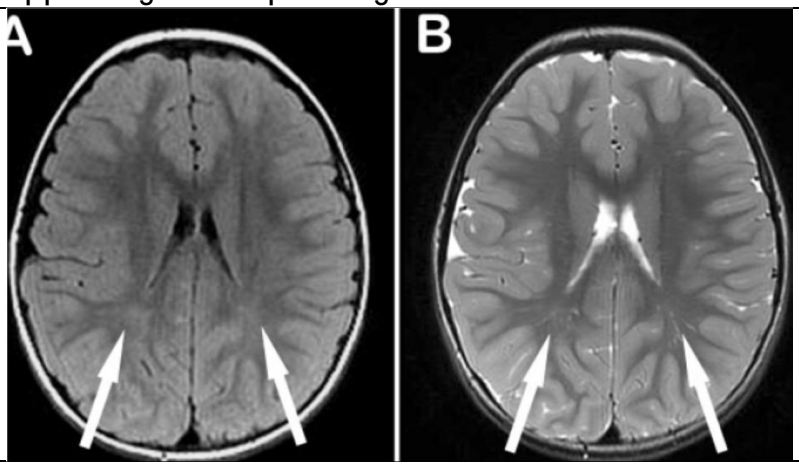
Complete myelination + Terminal zones of myelination



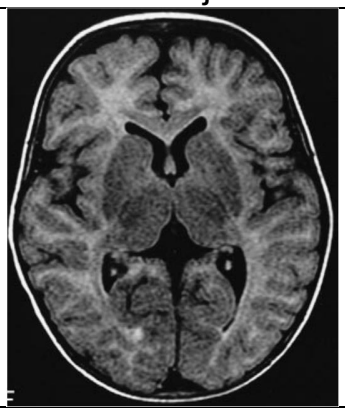
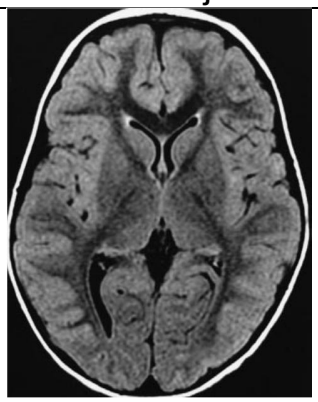


❖ Terminal Zone on Myelination:

- The brain myelination reaches the adult appearance at age of 2years.
- Persistent T2 hyperintensity beyond 2years of age at peritrigonal region. (DD: Periventricular leukomalacia PVL)
- No / faint hyperintense FLAIR signal
- No ventricular dilatation
- No WM volume loss
- Normal-appearing WM separating these zones from the ventricles



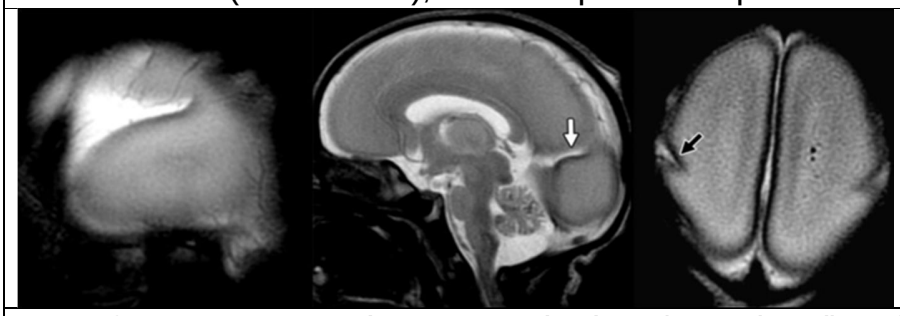
❖ FLAIR triphasic sequence of signal change of cerebral deep WM relative to adjacent GM

1st phase: At birth	2nd phase <1y	3rd phase >2y
Deep cerebral WM hypointense relative to adjacent GM	Deep cerebral WM hyperintense relative to adjacent GM	Deep cerebral WM hypointense relative to adjacent GM
		

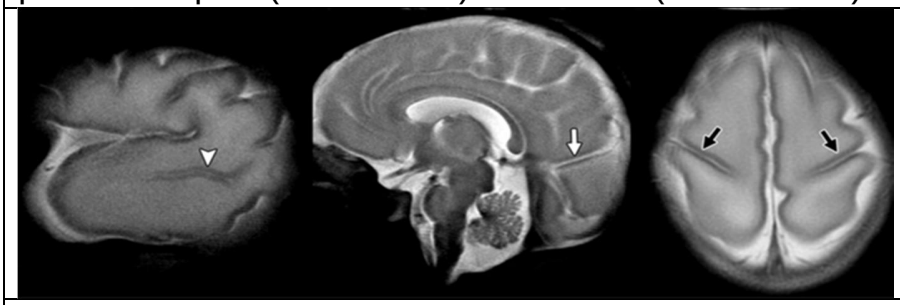
❖ Normal Sulcal Development

- With increasing age, the sulci become deeper, broader, and tortuous.
- By 34 weeks, all primary and most secondary sulci are present.

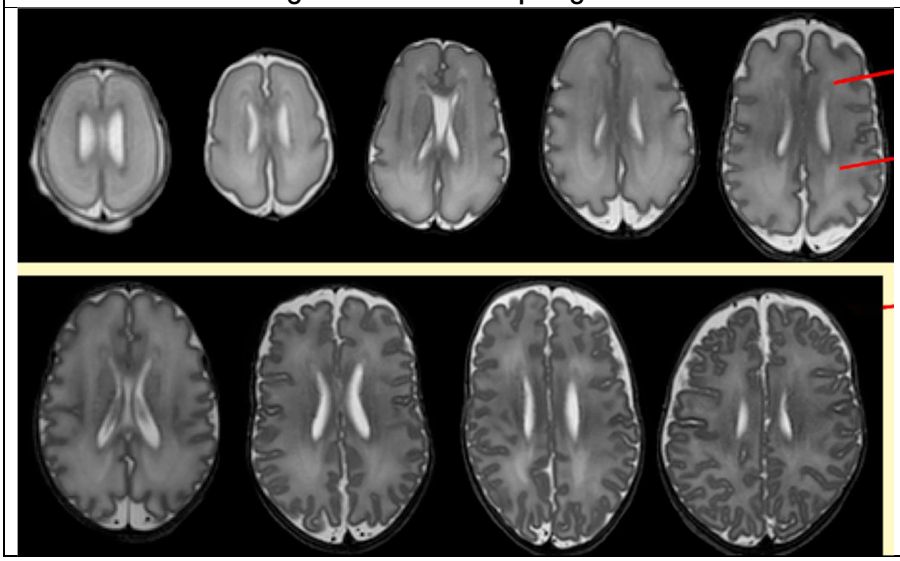
25w of gestation: Parieto-occipital sulcus (white arrow) and a rudimentary central sulcus (black arrow), but no superior temporal sulcus.



29w of gestation: superior temporal sulcus (arrowhead) and deepening of parieto-occipital (white arrow) and central (black arrows) sulci.



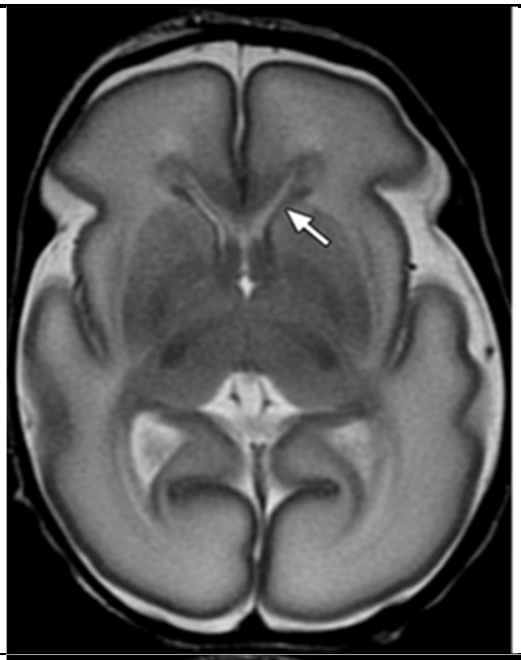
From 24w to 48w gestation show progressive increase in cortical folding



❖Migrational Milestones

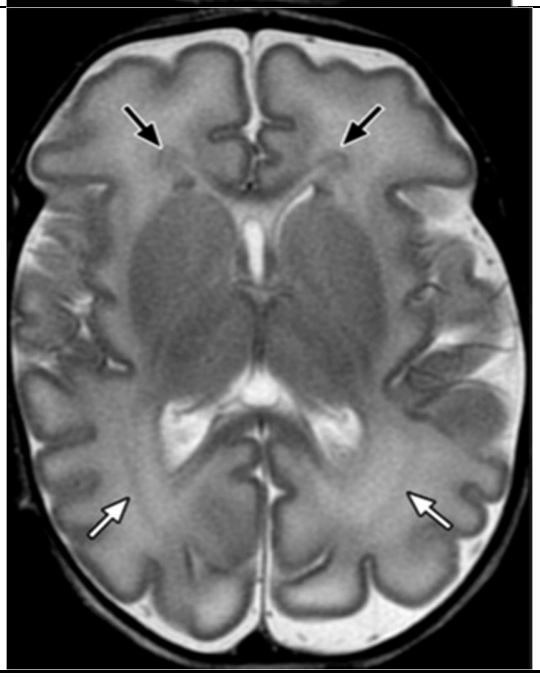
28–30 w gestational age:

- The germinal matrix is depicted on T2 as a low SI band that extends along the ventricular wall.
- The band is most prominent at the caudothalamic groove (ganglionic eminence).



Before 33 w gestational age:

- The white matter has a laminated appearance, with three alternating bands of different signal intensity on T2.
- Frontal horn “caps” and posterior periventricular “arrowheads” represent white matter “crossroads” containing dense extracellular matrix.



Leukodystrophy

- Dysmyelinating diseases, or leukodystrophies, encompass a wide spectrum of inherited neurodegenerative disorders affecting the integrity of myelin in the brain and peripheral nerves.

Classification of Leukodystrophies on the Basis of Organelle Disorder

Lysosomal storage diseases with white matter involvement
- Metachromatic leukodystrophy
- Krabbe disease (globoid cell leukodystrophy)
- Niemann-Pick disease
- Fabry disease
- GM_1 gangliosidosis
- GM_2 gangliosidosis
- Mucopolysaccharidosis
- Fucosidosis
- Mucolipidosis
- Wolmann disease and cholesterol ester storage disease
- Ceroid lipofuscinosis

Peroxisomal disorders
- Zellweger syndrome
- X-linked adrenoleukodystrophy (ALD)
- Neonatal ALD
- Pseudoneonatal ALD
- Classic Refsum disease
- Hyperpipecolic acidemia
- Cerebrotendinous xanthomatosis
- Abetalipoproteinemia
- Rhizomelic chondrodysplasia calcificans punctata

Mitochondrial dysfunction with leukoencephalopathy
- Leigh disease
- Myopathy, encephalopathy, lactic acidosis, and strokelike episodes (MELAS syndrome)
- Myoclonus epilepsy with ragged red fibers (MERRF syndrome)
- Kearns-Sayre syndrome

Disorders of amino acid and organic acid metabolism
- Canavan disease

White matter disorders with unknown metabolic defect
- Pelizaeus-Merzbacher disease (PMD)
- Alexander disease
- Congenital muscular dystrophy (Fukuyama type)

Simple approach for leukodystrophy:

- **Diffuse:**
  1- Diffuse with megalencephaly = Canavan.
  2- Diffuse with no megalencephaly & involving U fibers = Pelizaeus Merzbacher disease.
  3- Diffuse with no megalencephaly & sparing U fibers = Metachromatic leukodystrophy.

B. **Anterior:**
  1- Anterior with megalencephaly = Alexander.
  2- Anterior with no megalencephaly = metachromatic leukodystrophy.

C. **Posterior:**
  1- Posterior & confluent & enhanced = Adrenoleukodystrophy.
  2- Posterior & multifocal & non enhanced = Krabbe.

D. **Periventricular:**
  1- Confluent = Metachromatic leukodystrophy.
  2- Multifocal = Krabbe.

E. **Subcortical (U fibres):**
  1- Mitochondrial disorders = organic aciduria: Leigh & Kerne Sayre & glutaric aciduria).

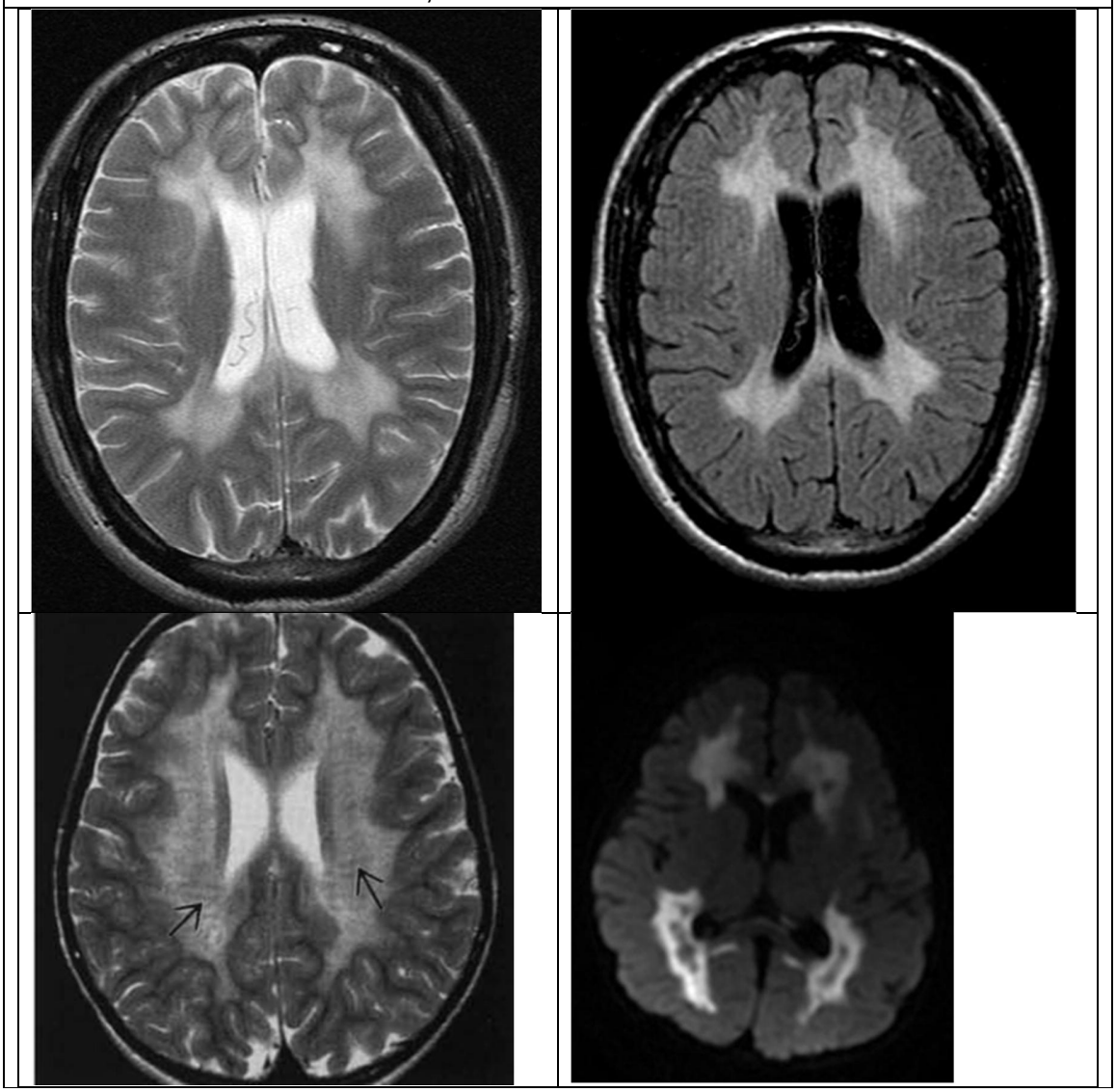
F. **Brain stem & Basal ganglia:**
  1- Diffuse brain stem involvement = Central pontine myelinolysis.
  - Focal brain stem involvement = Mitochondrial disorders (Organic aciduria = elevated lactate level on MRS) = MELAS & Glutaric aciduria & Leigh syndrome & Kearns Sayre syndrome.

A- Diffuse Pattern:

1) Metachromatic Leukodystrophy

- A lysosomal storage disorder
- Bilateral symmetrical confluent areas in periventricular WM
- "Tigroid pattern"
- Sparing subcortical U fibers.
- T1: low signal
- T2 /FLAIR: High signal
- +C: No contrast enhancement
- MRS: Elevation of choline, Maintained NAA.

Sulfatase enzyme

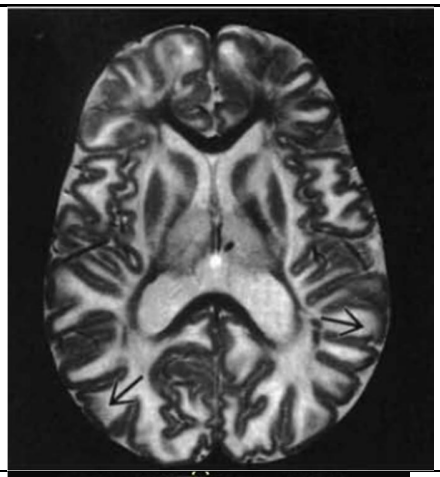


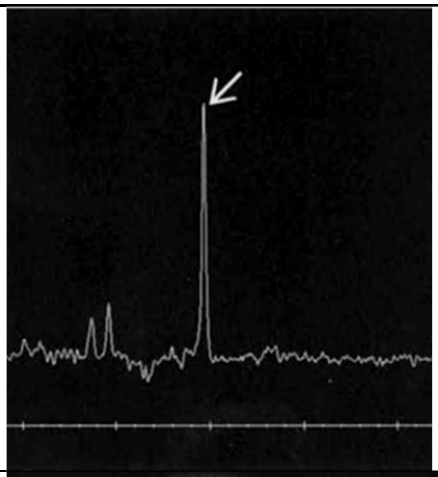
2) Canavan Disease

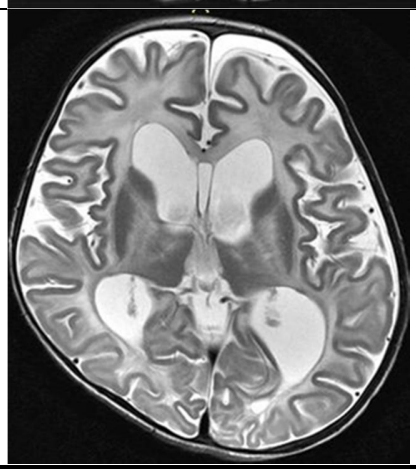
- Autosomal recessive disease

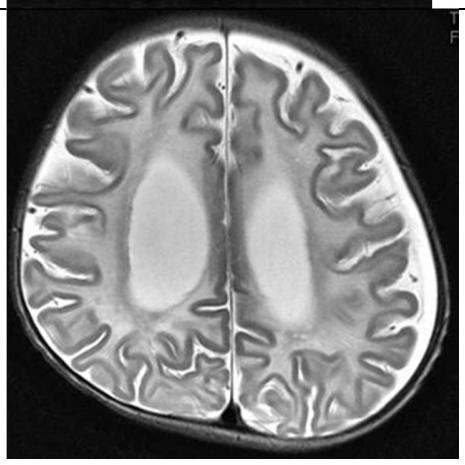
- Megalencephaly
- Diffuse bilateral involvement of WM & sub cortical U fibres.

- T1: low signal in white matter
- T2: high signal in white matter
- +C: NO enhancement
- MRS: Markedly elevated NAA and NAA/Cho ratio







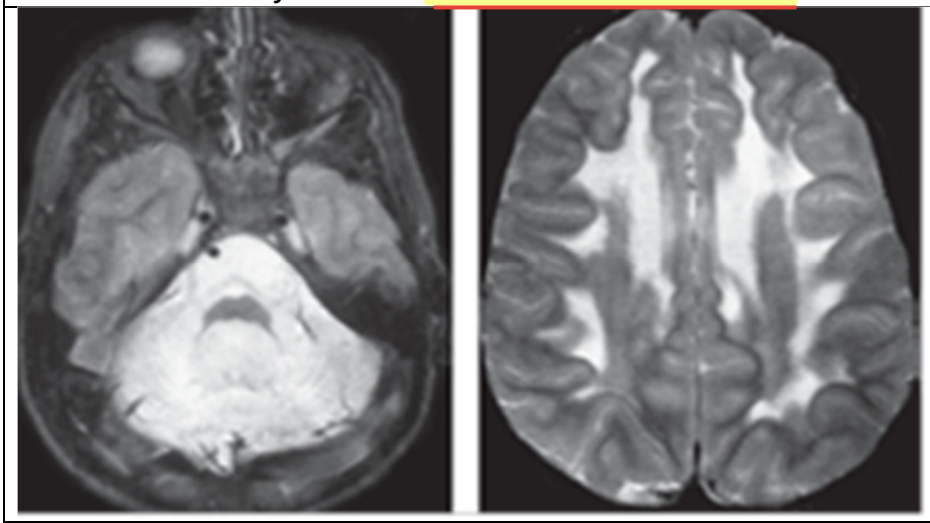


3) Pelizaeus-Merzbacher Disease

- X-linked recessive
- Hypotonia, respiratory distress, stridor, nystagmus, and profound myelin loss.

- No Megalencephaly
- Diffuse bilateral involvement of WM + sub cortical U fibres + Cerebellum.

- T1: low signal in white matter
- T2: high signal in white matter
- +C: NO enhancement
- MRS: Markedly elevated NAA and NAA/Cho ratio.

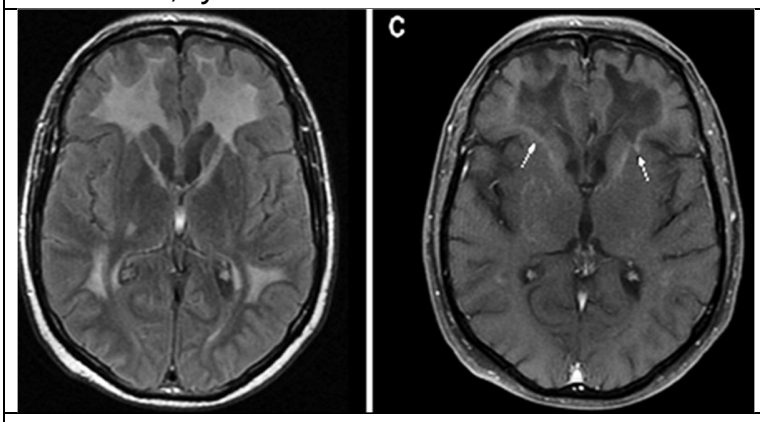


B- Anterior Pattern => Alexander Disease

3 clinical forms:

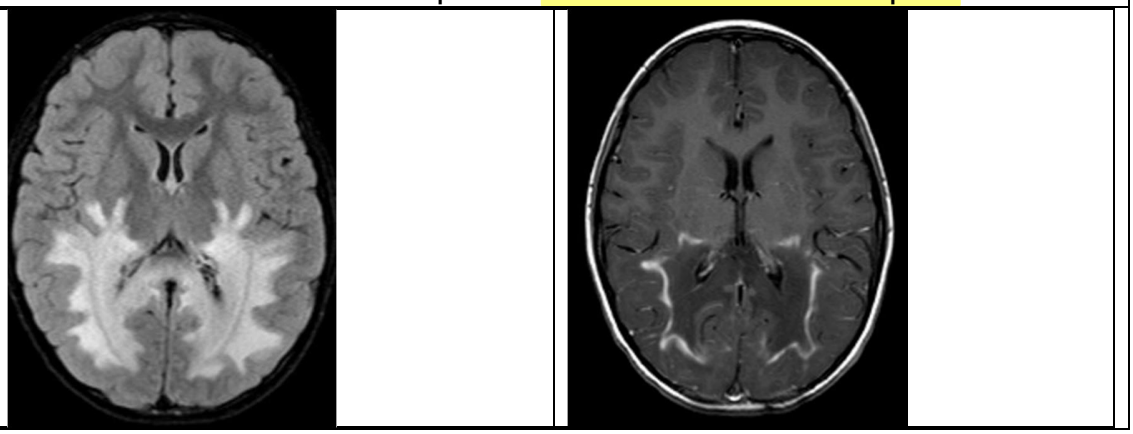
- Infantile / childhood onset
- Juvenile onset
- Adult onset (AOAD)

- Diffuse, symmetrical *bifrontal* abnormal WM signal and enhancement



C- Posterior Pattern => Adrenoleukodystrophy

- X linked inheritance.
- Symmetrical cerebral white matter signal change involving the posterior periventricular WM
- T1: hypointense
- T2/FLAIR: Markedly hyperintense
- +C: Garland-shaped enhancement may be visible in the anterior most periphery of the lesions.
- MRS: Decrease in the NAA peak + Elevation in the lactate peak



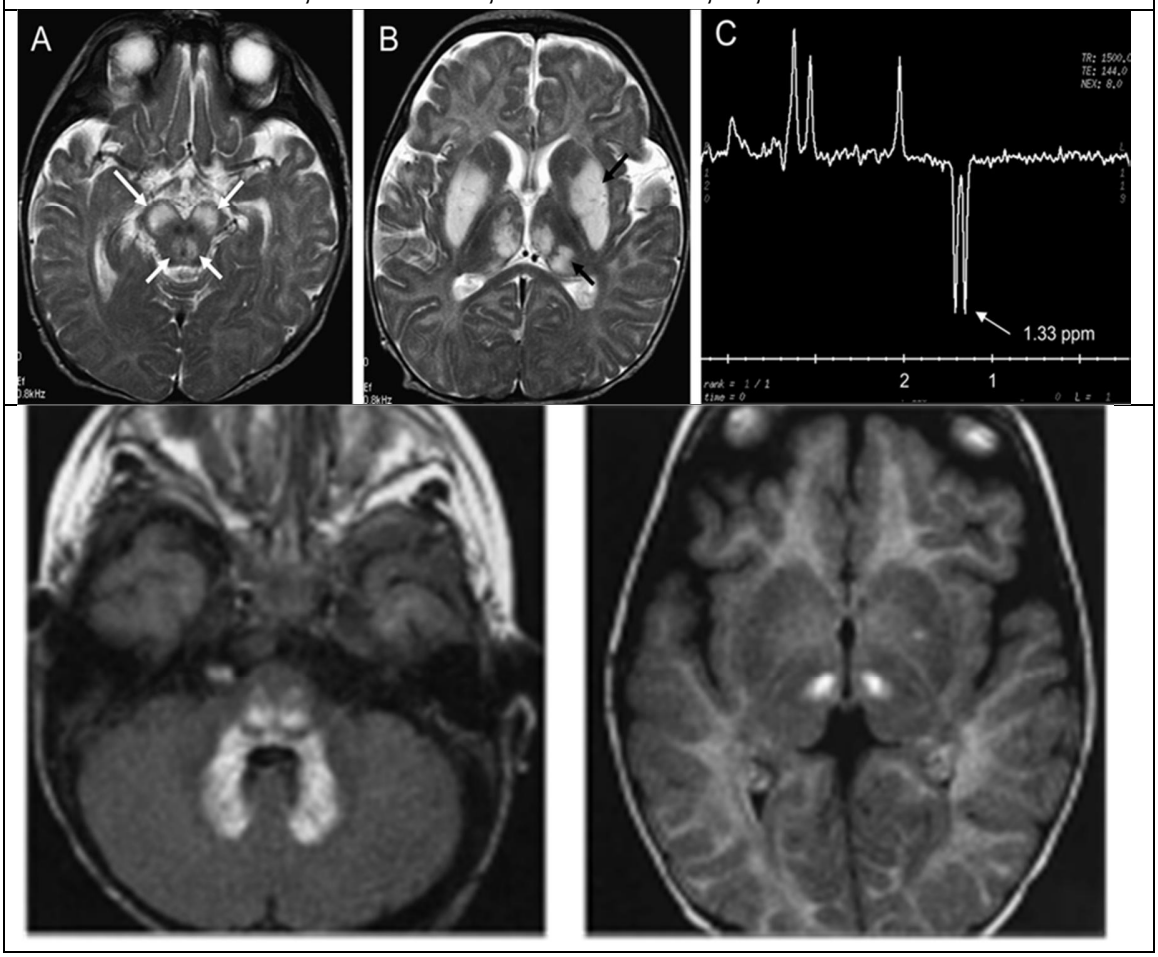
D- Basal Ganglia Pattern:

1) Leigh Syndrome

- Subacute necrotizing encephalomyelopathy (SNEM)) is a progressive neurodegenerative disorder

- Putamen, Midbrain, pons,
- Periaqueductal grey matter, substantia nigra,
- Floor of fourth ventricle
- Dentate nuclei.

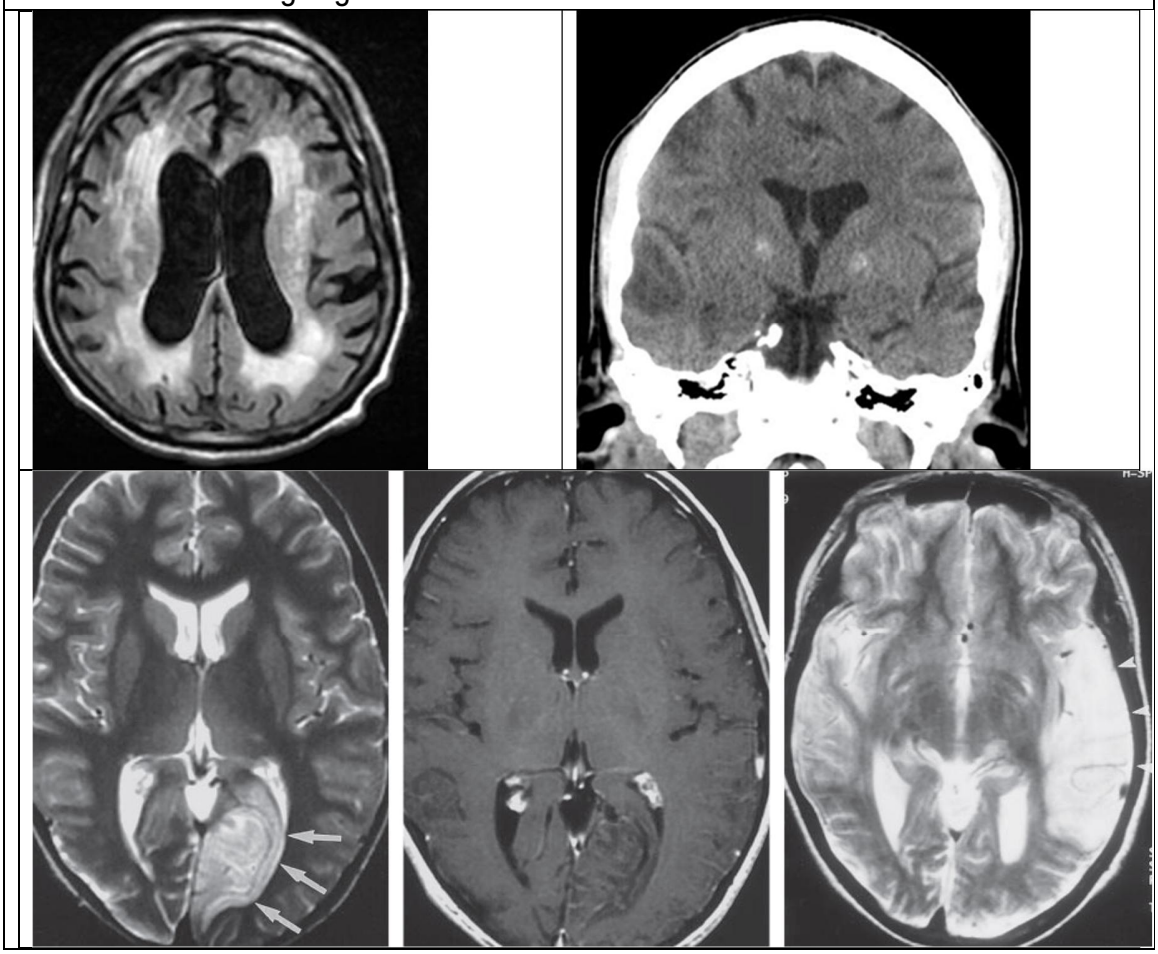
- T2/FLAIR: High signal
- MRS: ++ Lactate, Mild – NAA, NORMAL Cho, Cr,



2) MELAS

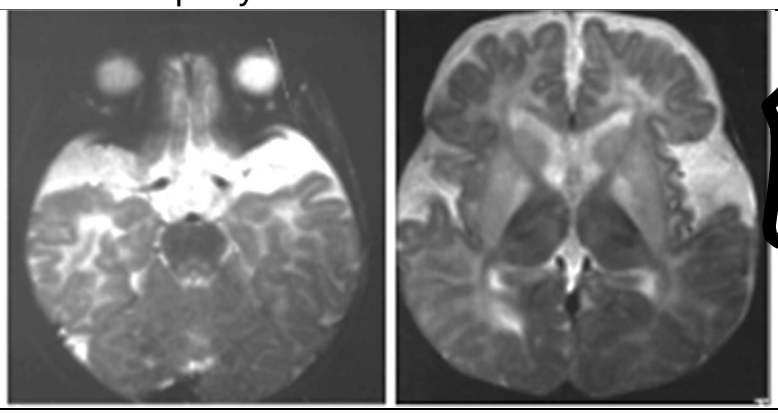
- Mitochondrial Encephalopathy With Lactic Acidosis And Stroke-like Episodes

- Multiple cortical and subcortical infarct-like lesions
- Parieto-occipital and parieto-temporal involvement is most common
- Bilateral basal ganglia calcification



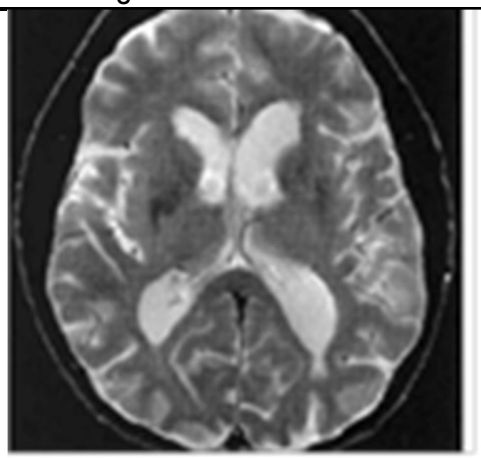
3) Glutaric Acidemia

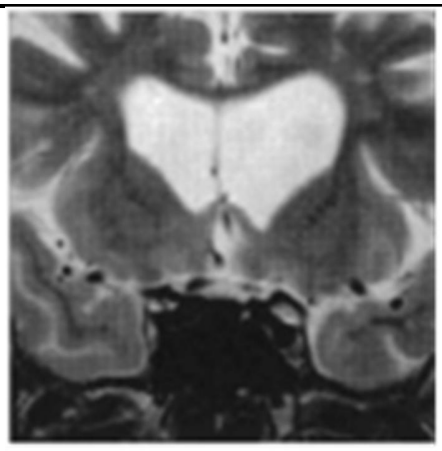
- Widening of sylvian fissures
- Bilateral basal ganglia lesions
- Macrocephaly



4) Huntington Disease (HD) = Huntington Chorea

- AD neurodegenerative disease with loss of GABAergic neurons of basal ganglia (BG)
- Clinical triad: Early onset dementia, choreoathetosis, and psychosis
- Adult-onset HD = 35-44 y
- Juvenile HD (5-10% of cases): < 20 Y
- Atrophy of caudate nucleus (Cn)
- Enlargement of frontal horns

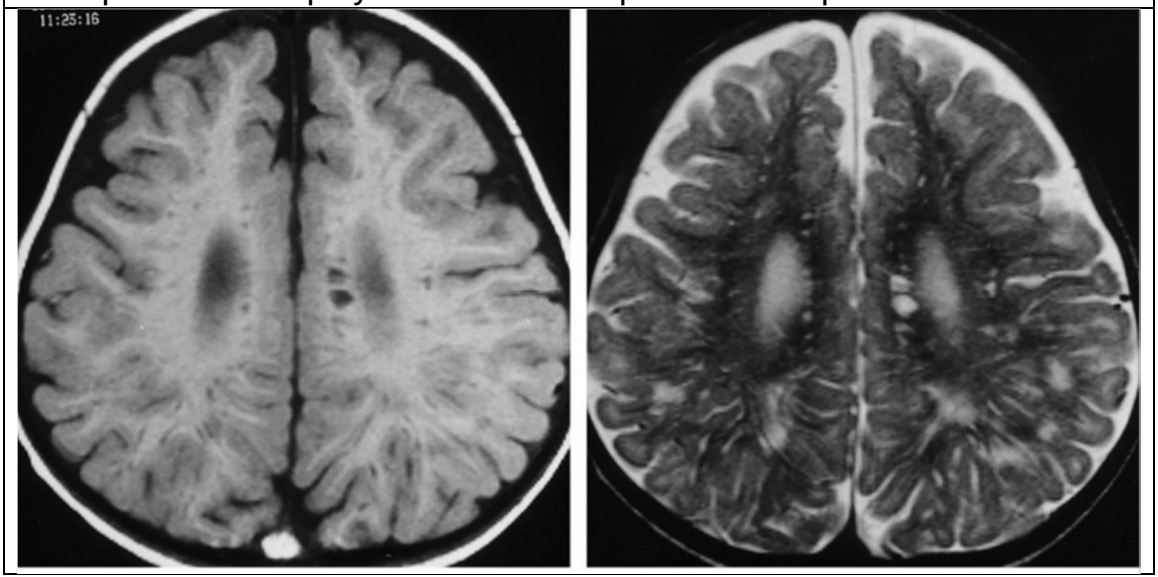




Mucopolysaccharidosis

- Mucopolysaccharidosis is caused by a deficiency of the various lysosomal enzymes involved in the degradation of glycosaminoglycans

- Multiple sharply defined foci are commonly present in the corpus callosum, basal ganglia, and cerebral white matter.
- CSF like signal
- Represent mucopolysaccharide-filled perivascular spaces



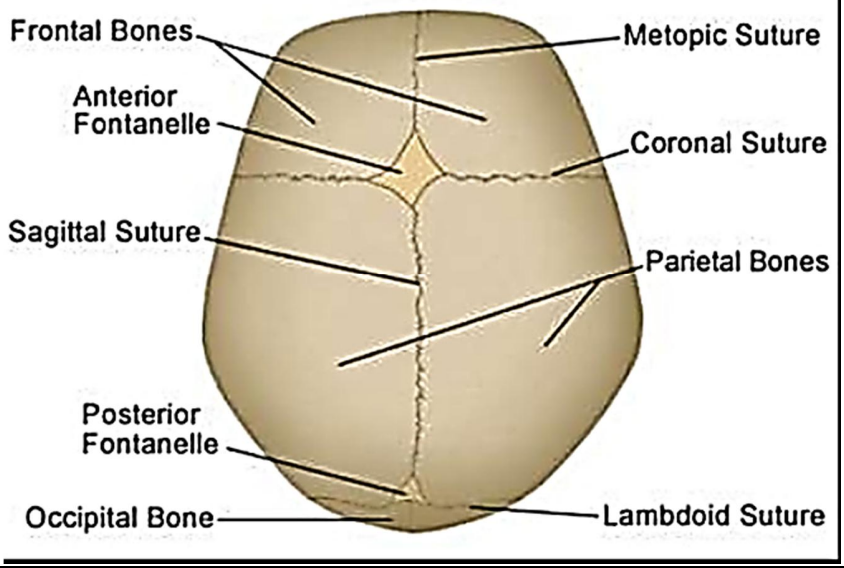
Microcephaly

Microcephaly = Small head size
Head circumference > 3 SD below the mean for age and sex
• ↓ Craniofacial proportions • Normal craniofacial ratios: Premature (5:1), term (4:1),2 yrs (3:1), 3 yrs (2.5:1), 12 yrs (2:1), adult (1.5:1)
<u>Primary (Genetic):</u> • Small, but grossly normal brain • Gyral simplification • or ± pachygyric, lissencephalic, holoprosencephalic • or ± hypoplastic cerebellum, hypomyelination
<u>Secondary (Nongenetic):</u> • Results from noxious agent that affects fetal/infant brain growth • Hypoxic ischemic encephalopathy (HIE) • TORCH infection • Fetal alcohol syndrome (FAS): Callosal abnormalities, ventriculomegaly • Nonaccidental head injury (NAHI): Encephalomalacia, chronic subdurals.

Craniosynostosis

Premature fusion (osseous obliteration) of cranial sutures

Normal Skull of Infant



Age of fusion

• Metopic suture = 3m-2y • Lambdoid, sagittal and coronal: 40y • Squamosal = 40 -70y	• Posterior fontanelle = 3m • Sphenoidal = 6m • Mastoid = 6-18m • Anterior = 1-3y
15% Syndromatic 85% Non-syndromatic	75% Simple 25% Multi-suture

Scaphocephaly	50-60%
Frontal Plagiocephaly	20-30%
Turribrachycephaly	10-15%
Trigonocephaly	5-10%
Posterior Plagiocephaly	2-4%

1ry Signs Of Craniosynostosis

- Absence of suture
- Bony bridging along the suture
- Heaping up of bone at the suture
- Sutural narrowing

2ry Signs Of Craniosynostosis

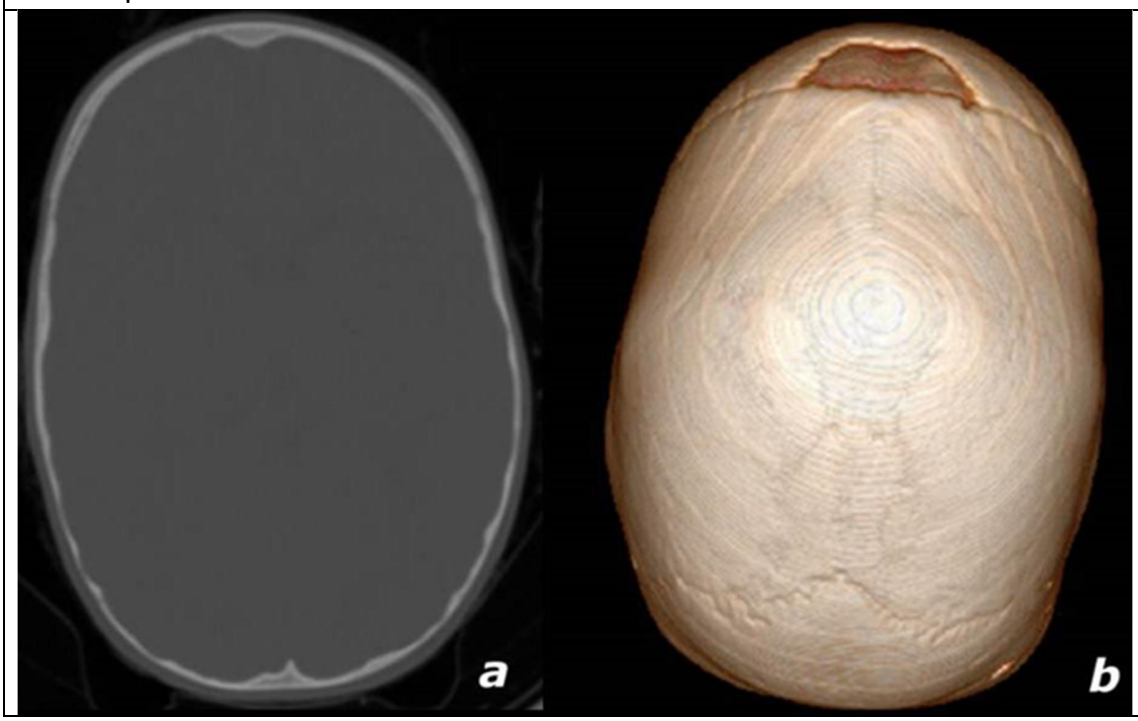
- Altered calvarial shape
- ↑ Calvarial growth parallel to the fused suture
- ↓ Calvarial growth perpendicular to the fused suture
- Facial and orbital anomalies.
- Effacement of the underlying subarachnoid spaces
- Restriction on underlying brain growth, and is considered an indication for surgical repair

CT with 3D image reformations is the best modality used for evaluation of sutures.

A- Single-Suture Synostosis (Primary)

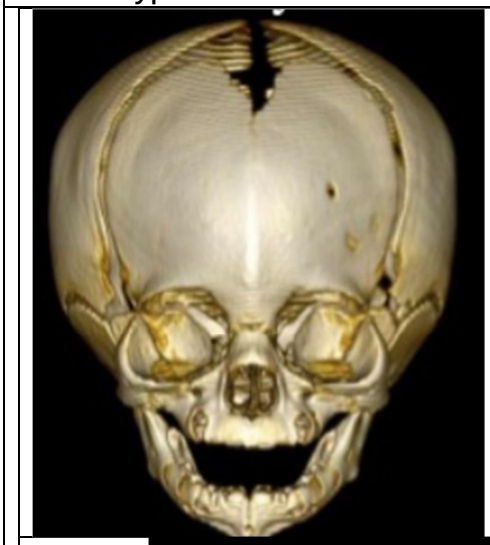
1) Scaphocephaly (Dolichocephaly) = Sagittal synostosis

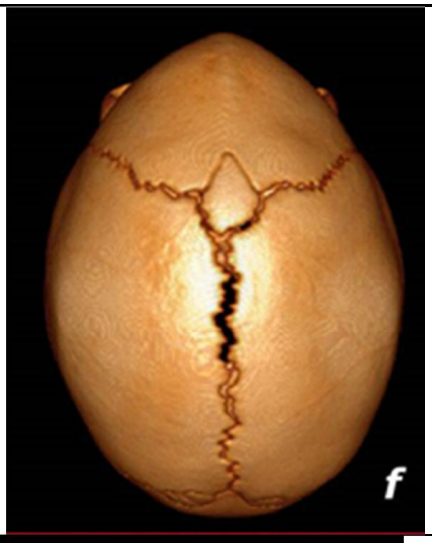
- Most common (50 – 60%)
- ↑ AP + ↓ Transverse

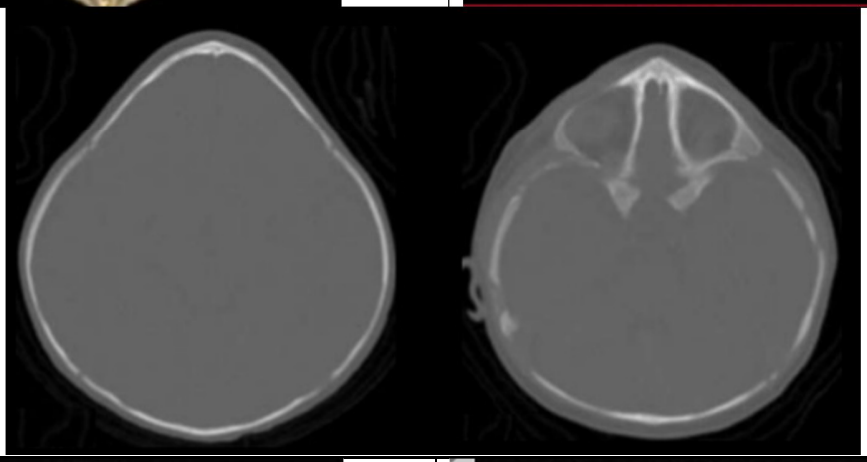


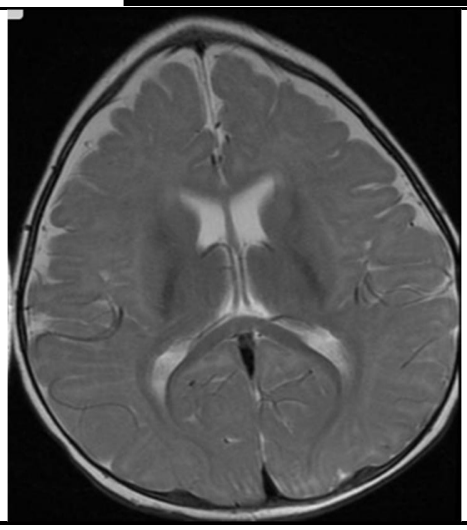
2) Trigonocephaly = Metopic synostosis

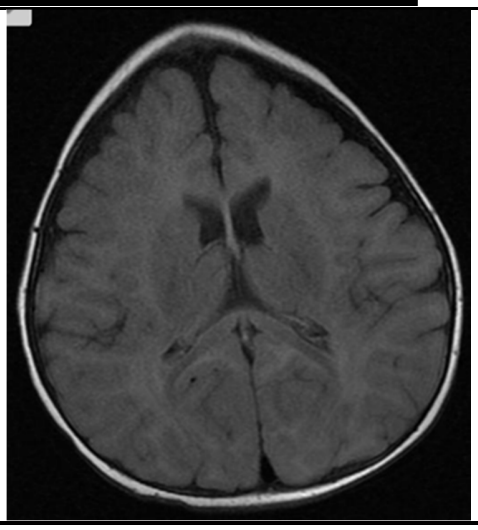
- "Ax-head" - "pear-shaped" or triangle shaped head
- Hypotelorism





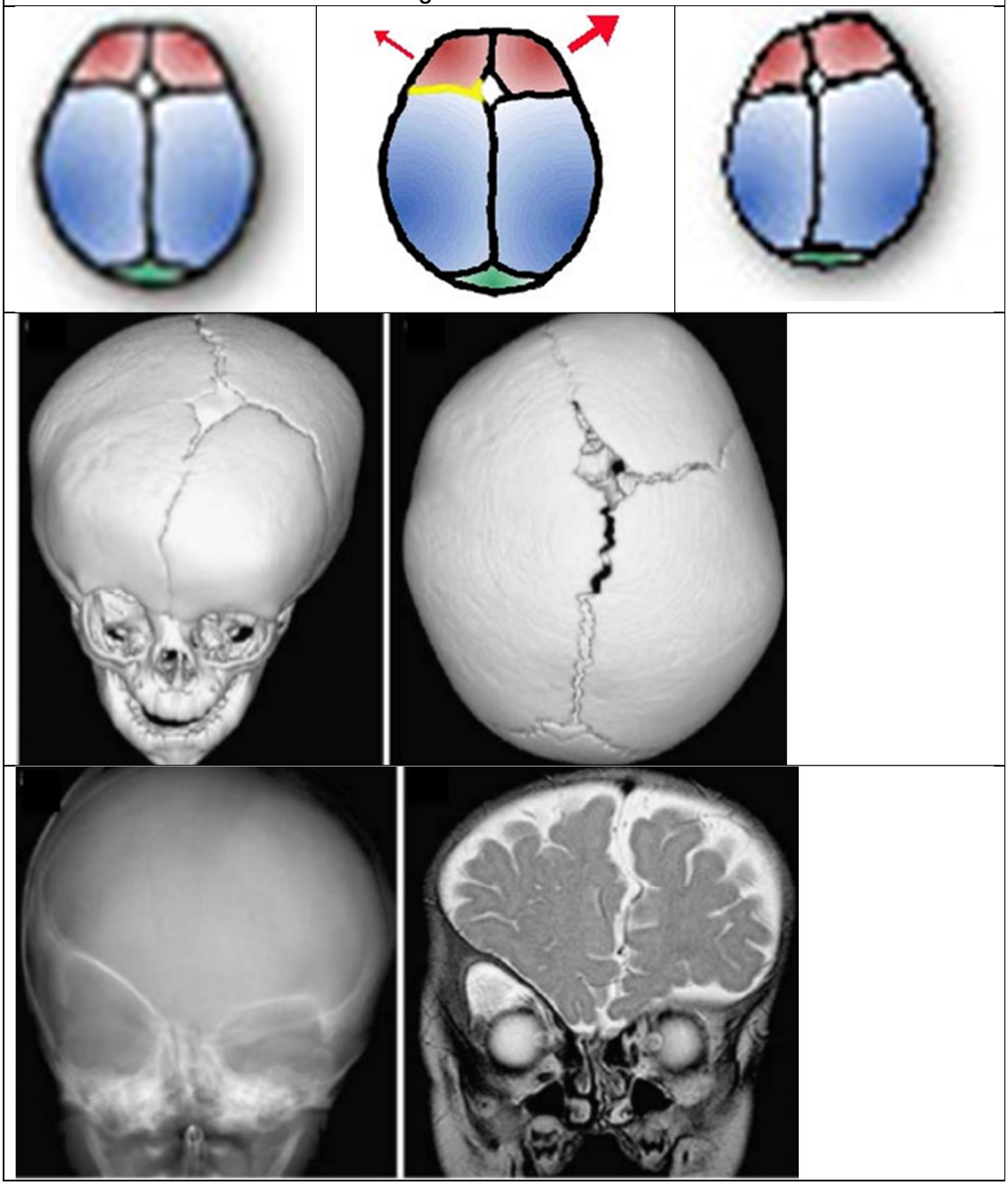






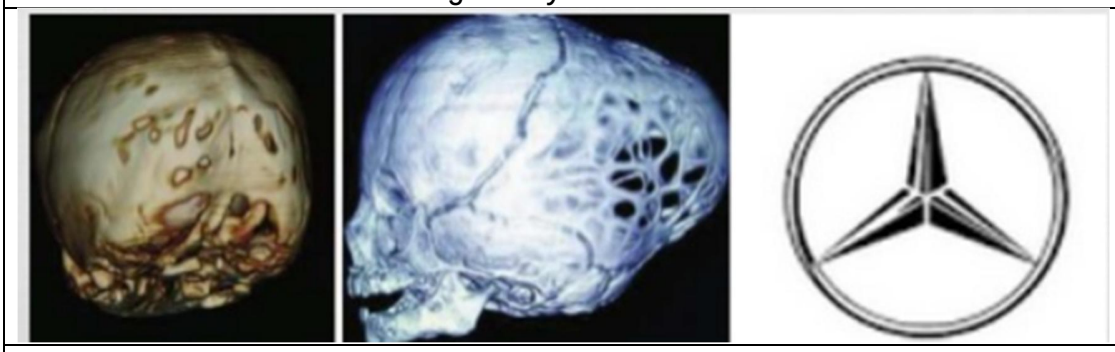
3) Anterior / Frontal Plagiocephaly = Unilateral Coronal Synostosis

- Bent head
- Ipsilateral frontal flattening
- Ipsilateral Harlequin orbit
- Contralateral frontal bossing



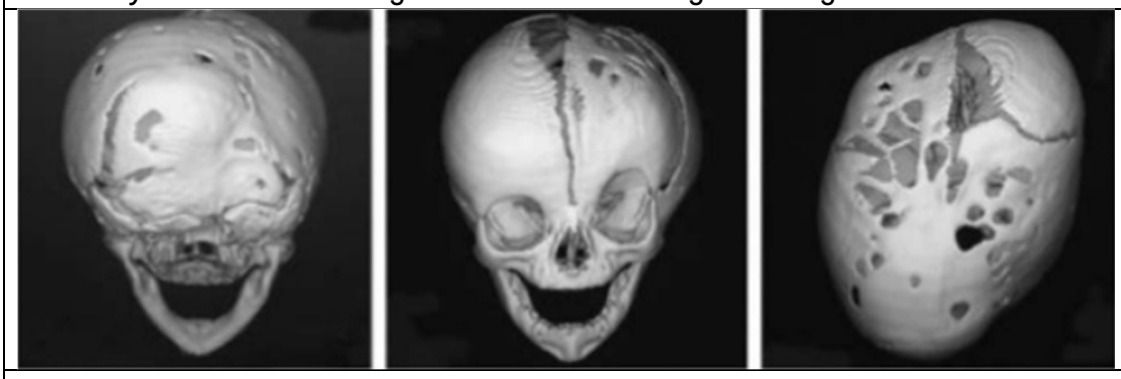
3) Mercedes-Benz pattern:

- Bilateral lambdoid + sagittal synostosis



4) Z-pattern:

Craniosynostosis involving the left cornal + sagittal + right lambdoid sutures

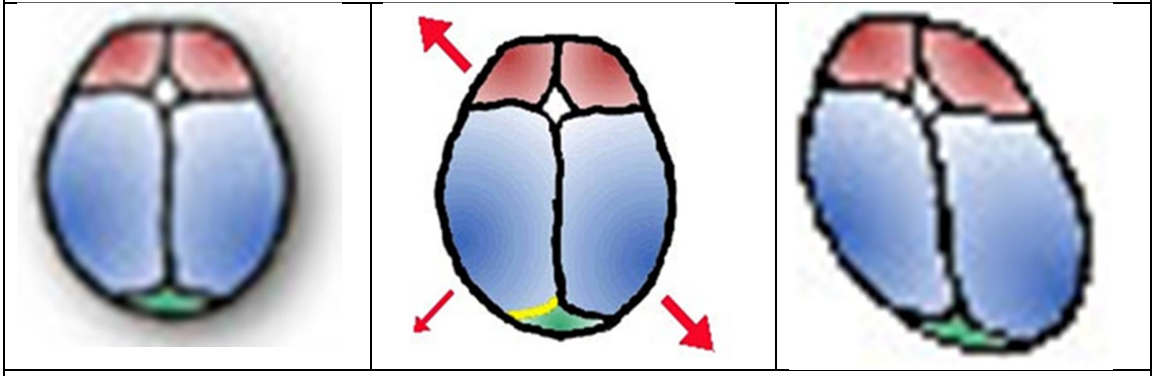


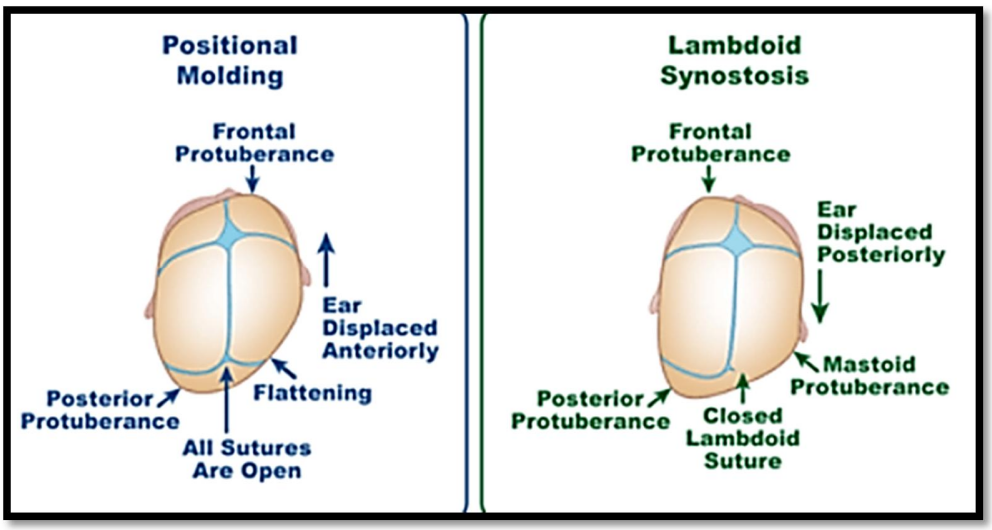
D- Syndromic Craniosynostosis

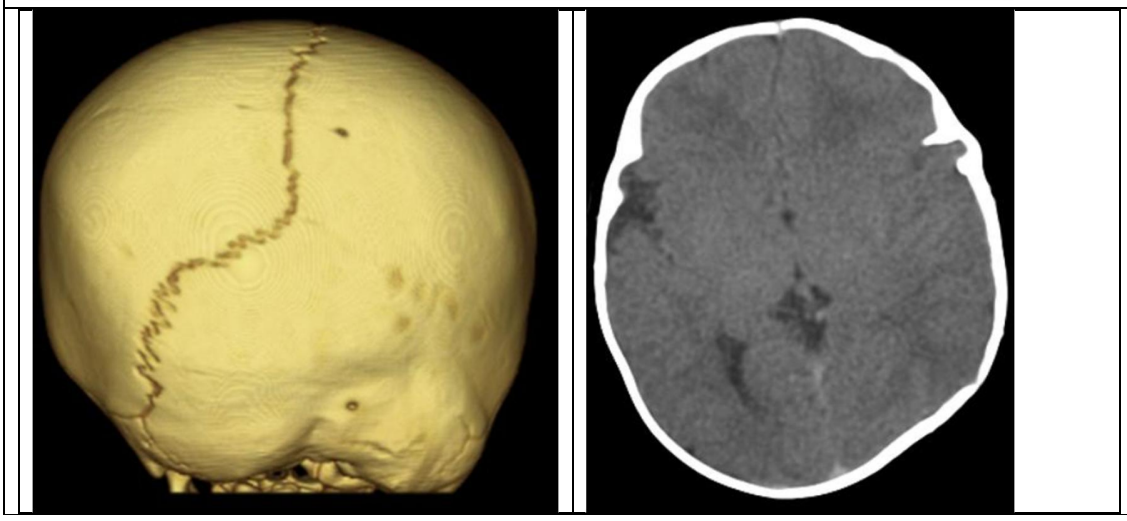
- Acrocephalosyndactyly/acrocephalopolysyndactly types
  - Apert syndrome
  - Carpenter syndrome
  - Crouzon syndrome
- Choanal atresia

4) Posterior / Occipital Plagiocephaly = Unilateral Lambdoid Synostosis

- Slant-Head
- Ipsilateral occipital flattening
- Contralateral occipital prominence







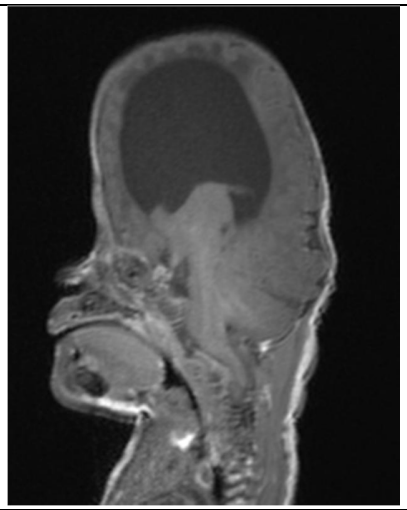
B- Double-Suture Synostosis

1) Bicoronal: Skull is wider than normal (Anterior brachycephaly)
2) Bilambdoid: Skull is wider than normal (Posterior brachycephaly)
3) Sagittal plus metopic: Head is long and narrow (Scaphocephaly)

C- Complex Multisuture Synostosis

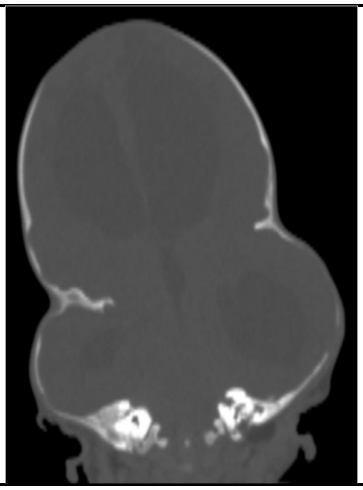
1) Oxycephaly

- Fusion of all skull sutures and basilar skull sutures
- Tower-like skull



2) Kleeblattschadel:

- Fusion of all skull sutures (sagittal, coronal and lambdoid sutures) except metopic and squamosal
- Cloverleaf skull



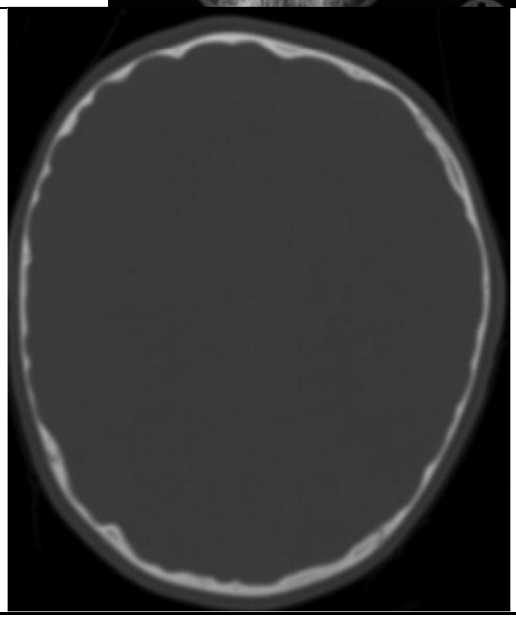
E- Secondary Nonsyndromatic Craniosynostosis

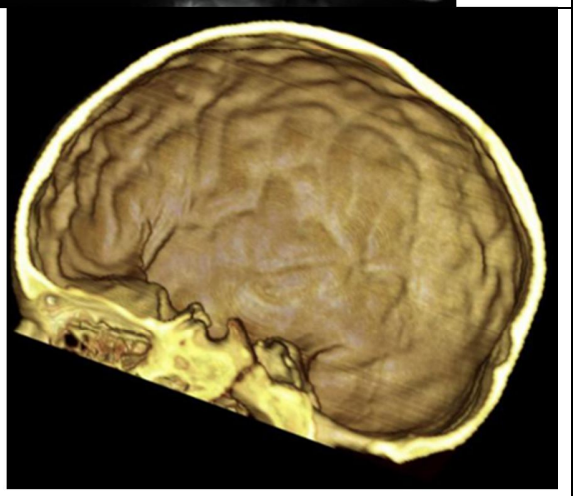
- Metabolic Disorders
  - Rickets
  - Hyperthyroidism
- Hematological Disorders
  - Thalassemias
  - Sickle cell anemia
  - Polycythemia vera
- Lysosomal Storage Disorders
  - Sly Syndrome
  - Hurler Syndrome
  - Morquio Syndrome
- Teratogens
  - Phenytoin
  - Valproic Acid
  - Retinoic Acid
  - Aminopterin
- Malformations
  - Microcephaly
  - Encephalocele
  - Shunted Hydrocephalus
- Positional Etiology
  - Fetal Head Restraints
  - Deformational Plagiocephaly

❖Copper Beaten Skull

- Refers to the prominence of convolutional markings (gyral impressions on the inner table of the skull) seen throughout the skull vault.

- Raised intracranial pressure is the causative mechanism behind increased inner table markings that give rise to the copper beaten skull appearance:
  - → Craniosynostosis
  - → Obstructive hydrocephalus
  - → Hypophosphatasia
  - → Intracranial masses

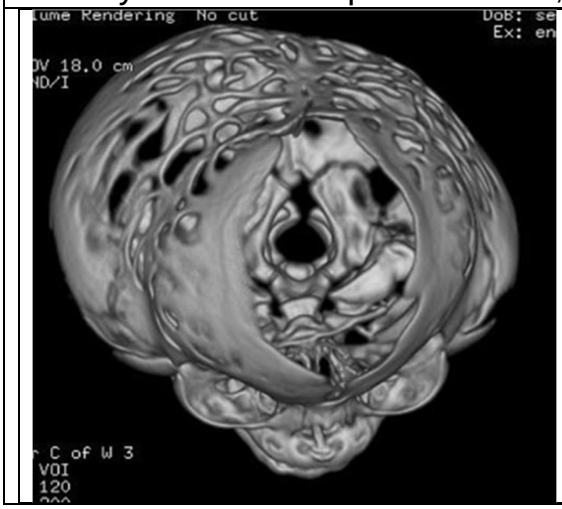




❖ Luckenschadel Skull

- Also known as lacunar skull or craniolacunae, is a dysplasia of the membranous skull vault and is associated with Chiari II malformations (seen in up to 80% of such cases).
- The inner table is more affected than the outer, with regions of apparent thinning (corresponding to nonossified fibrous bone) of the skull vault.
- The condition is self limiting, typically resolving after 6 months of age.

- Characterized by groups of round, oval or finger shaped pits on the inner surface of the vault (membranous part), separated by ridges of bone.
- They lie in thickest part of frontal, parietal and upper occipital bone.



Phakomatoses

A. Neurofibromatosis Type I
B. Neurofibromatosis Type II
C. Tubers Sclerosis
D. Von Hippel Lindau Syndrome
E. Sturge Weber Syndrome
F. Basal Cell Naevus Syndrome (Gorlin-Goltz Syndrome)
G. Wyburn-Mason Syndrome (Bonnet-Dechaume-Blanc Syndrome)
H. Encephalocraniocutaneous Lipomatosis
I. Hypomelanosis Of Ito
J. Nijmegen Breakage Syndrome
K. Epidermal Naevus Syndrome

- Phakomatoses are a group of neurocutaneous disorders characterized by involvement of structures that arise from the embryonic ectoderm (thus central nervous system, skin and eyes).
- Other organs may also be involved

A- Neurofibromatosis Type 1

- Von Recklinghausen disease
- Multisystem neurocutaneous disorder
- Most common phakomatosis.
- Most common inherited CNS disorders
- Autosomal dominant disorders
- 50% inherited

Clinical diagnosis ≥ 2 of following are required:

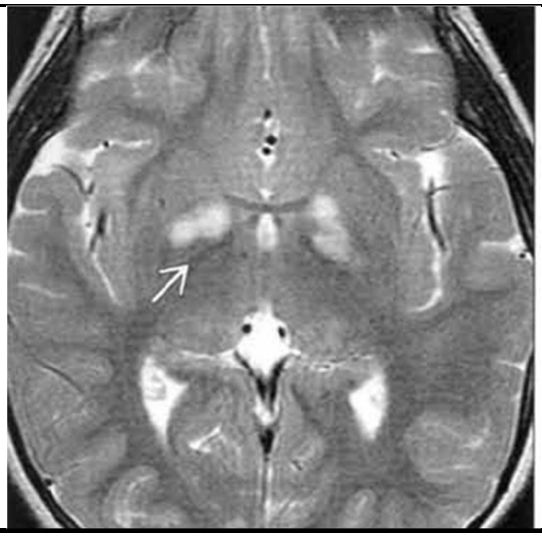
- ≥ 6 cafe au lait spots evident during one year
- ≥ 2 neurofibromas or 1 plexiform neurofibroma
- Optic nerve glioma
- Distinctive osseous lesion
- Sphenoid wing dysplasia
- Two or more iris hamartomas (Lisch nodules)
- Axillary or inguinal freckling
- Primary relative with NF-1

CNS Manifestations of NF-I

- FASI: focal areas of signal intensity in deep white matter and basal ganglia or corpus callosum i.e. areas of T2/FLAIR hyperintensity with no contrast enhancement
- Optic nerve glioma or optic pathway glioma
- Sphenoid wing dysplasia + Arachnoid cyst
- Buphthalmos
- Lambdoid suture defects
- Dural calcification at the vertex
- Moya-moya phenomenon (rare)

FASI:

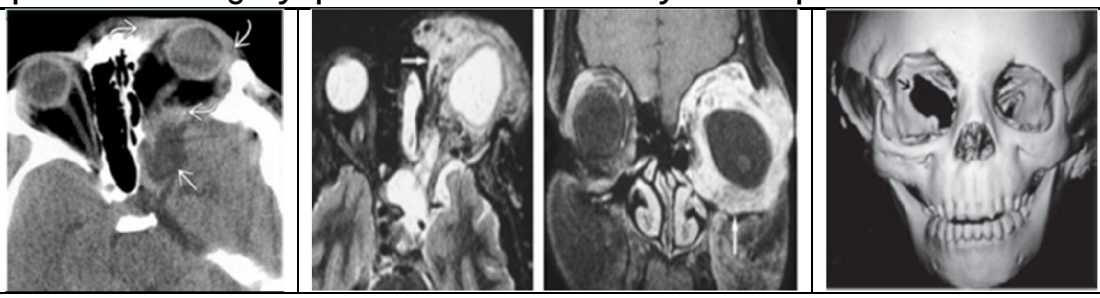
- Focal areas of signal intensity in deep white matter and basal ganglia or corpus callosum
- T2/FLAIR: hyperintensity
- +C: no enhancement



Optic Nerve Glioma

Plexiform Neurofibromatosis

Sphenoid wing dysplasia + Arachnoid cyst + Buphthalmos



B- Neurofibromatosis Type 2

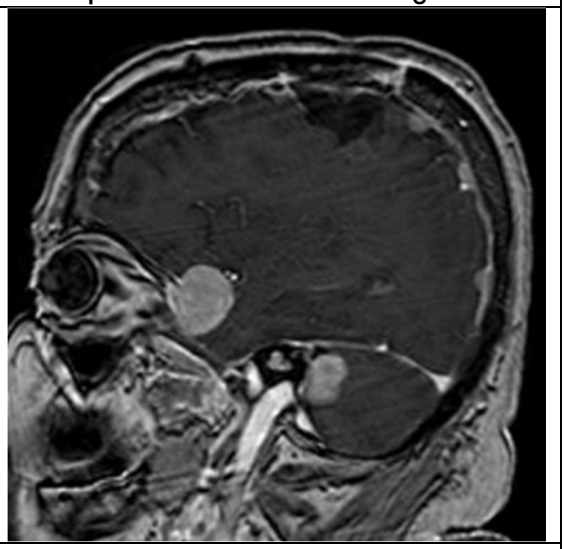
A rare autosomal dominant neurocutaneous disorder (phakomatosis) manifesting as development of multiple CNS tumours

MISME

Multiple Inherited Schwannomas, Meningiomas and Ependymomas

Bilateral Acoustic Schwannoma

Multiple intracranial meningiomas



Multiple intraspinal meningiomas + Schwannoma + Ependymoma

C- TUBEROUS SCLEROSIS

- Rare, Autosomal dominant (20%-50%) or sporadic (50%) or inherited (50%), neuroectodermal disorder.
- TS has been considered to be caused by mutations of two genes known as TSC1 and TSC2.

Major Features	Minor Features
Frequent	**Frequent**
Facial angiofibromas	Multiple pits in dental enamel
Hypomelanotic macules	Hamartomatous rectal polyps
Cortical tubers	
Subependymal nodules	
Common	**Common**
Retinal hamartomas	Bone cysts
LAM	Cerebral white matter radial migration lines
Renal AML	Multiple renal cysts
Cardiac rhabdomyomas	Gingival fibromas
Uncommon	**Uncommon**
Shagreen patches	"Confetti" skin lesions
Ungual fibromas	Retinal achromatic patches
Subependymal giant-cell tumors	

Definite diagnosis: two major features or one major feature plus two minor features
Probable diagnosis: one major feature plus one minor feature
Possible diagnosis: one major feature or two or more minor features

Age (y)	Main Causes of Death
0–9	Cardiovascular lesions, status epilepticus, bronchopneumonia
10–19	Brain tumors, renal failure, status epilepticus
20–29	Status epilepticus, renal failure, brain tumors
30–39	Renal failure, brain tumors, status epilepticus
40–49	Renal failure, LAM
>50	Renal failure, LAM

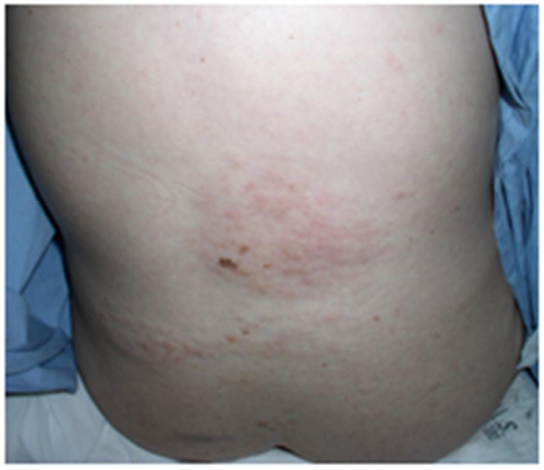
The clinical triad:

- Adenoma sebacum
- Seizures
- Mental retardation

A- Dermatological features:

Tuberous sclerosis

Adenoma sebaceum



Shagreen patch

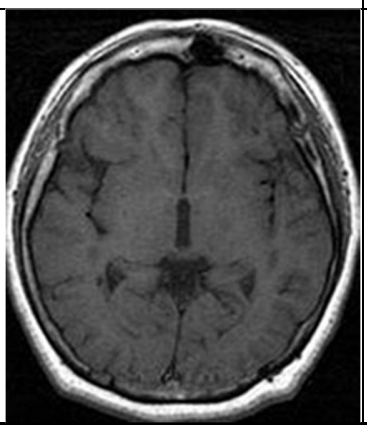
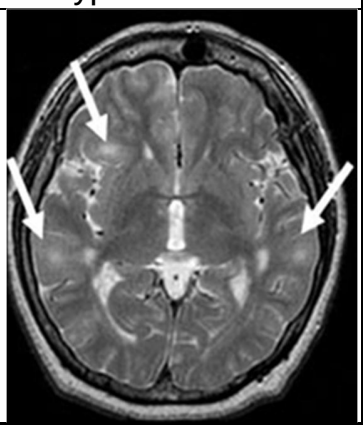
Ungual fibromas

B- CNS Involvement (4 major CNS lesions)

1. Cortical/subcortical tubers
2. Subependymal hamartoma (95-100%)
3. Subependymal giant cell astrocytoma (SGCA)
4. White matter lesions (40-90%)

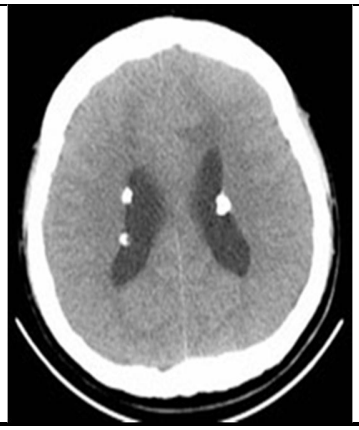
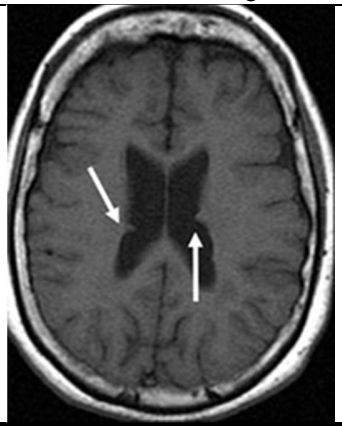
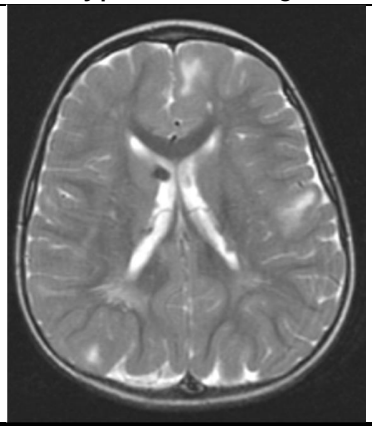
1) Cortical/subcortical tubers

- Multiple cortical and subcortical lesions
- Location: Supra>>>> infra tentorial
- Frontal> parietal> occipital> temporal> cerebellum
- CT : Ca^{++} with age
- T1+C: 10% of cortical tubers show enhancement
- MRS: Normal
- PWI: Hypovascular to WM

Axial T1:	Axial T2:	Axial FLAIR:
• The peripheral component is isointense to GM • The inner component is isointense to WM	• The peripheral component is isointense to GM • The inner component is hyper to WM	• The peripheral component is isointense to GM • The inner component is hyper to WM
		

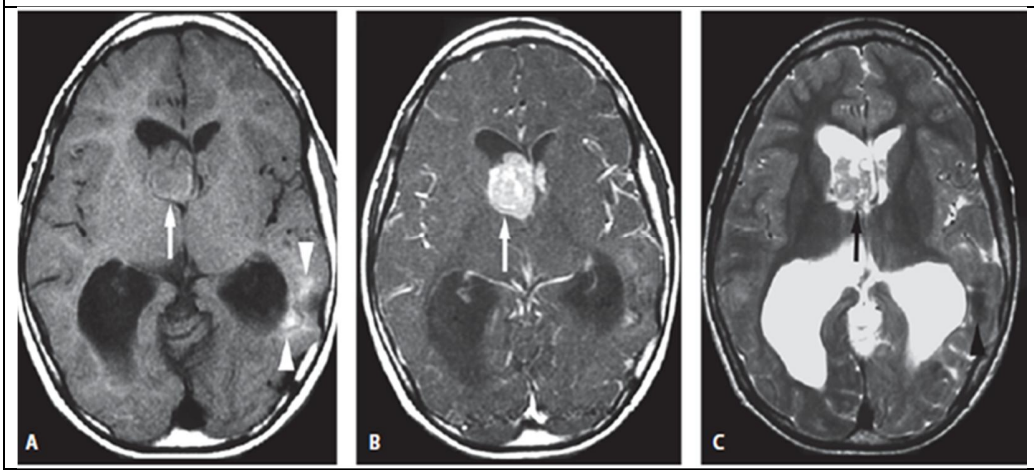
2) Subependymal Hamartoma

- Multiple subependymal nodules of 1-12mm
- CT: Ca ++ with age
- +C: May show minimal contrast enhancement

	T1: Isointense signal	T2: Hypointense signal
		

3) Subependymal Giant Cell Astrocytoma (SGCA)

- 15%
- WHO grade I
- Location: centered at foramen of Monro
- Size: Large > 1.2cm. Progressively enlarged
- Grow towards the ventricle not brain parenchyma
- Growth towards brain parenchyma suggests transformation to high grade.
- +C: Enhance



4) White Matter Lesions

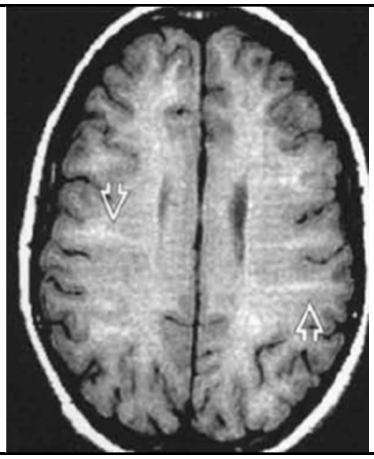
i. Superficial white matter abnormalities associated with cortical tubers
ii. Radial white matter bands
iii. Cyst like white matter lesions.

i. Superficial white matter abnormalities

- They are related to almost all cortical tubers.
- They reflect reduced myelin or increased gliotic reaction.
- T2: high-intensity areas
- T1: decreased-intensity areas

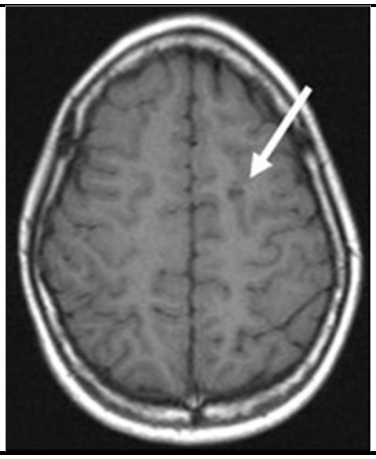
ii. Radial white matter bands:

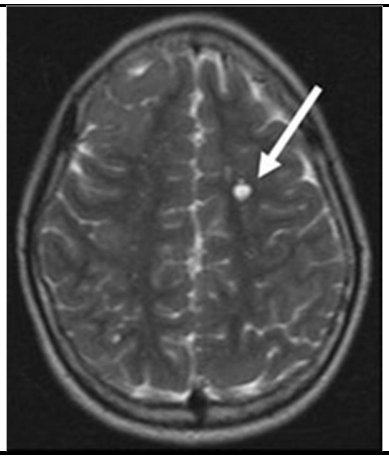
- Thin straight or curvilinear bands
- Run from ventricular or juxtaventricular white matter
- to the deep surface of cortical tubers or normal-appearing cortex



iii. Cyst-like white matter lesions (cystoid brain Degeneration)

- Small well-demarcated lesion of CSF like signal in all sequences are seen in white matter

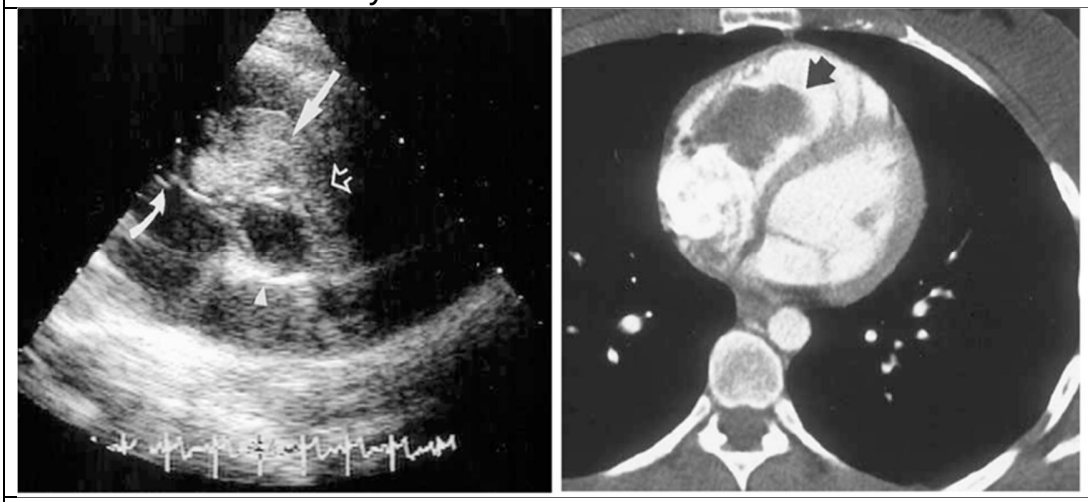




C- Cardiac Involvement

Cardiac Rhabdomyoma:

- Occur in 50-70% of pts with TS
- These harmatomas are usually multiple and intramural
- Most cardiac rhabdomyomas regress before birth.
- Spontaneous regression or disappearance of the tumor can occur in 70% of children by the age of 4 years.
- More than 80% of the tumors cause no clinical manifestations at birth.
- Only minority of cases cause abnormal valve function, outflow obstruction and arrhythmias



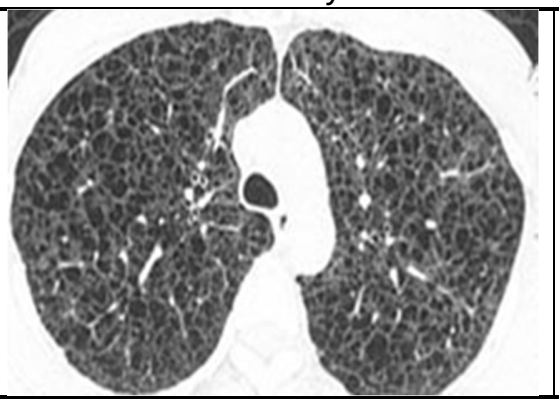
D- Eye Involvement

- Retinal hamartoma.
- Both eyes, multiple.
- May calcify: D.D: Retinoblastoma.

E- Pulmonary Involvement

Lymphangioleiomyomatosis (LAM):

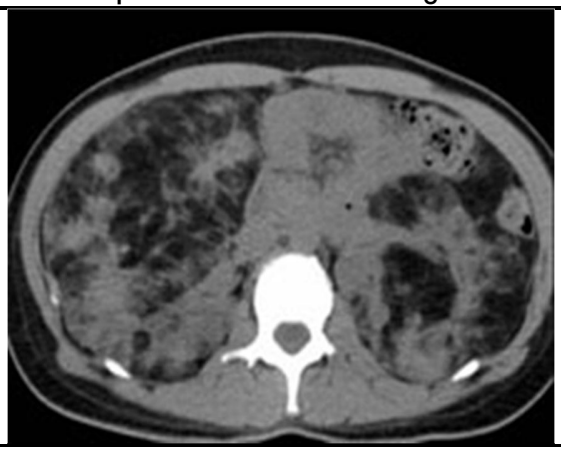
- Round, thin-walled cysts of variable size and contour
- Distribution: Symmetric and uniform throughout the lungs.
- Pneumothorax - Chylothorax

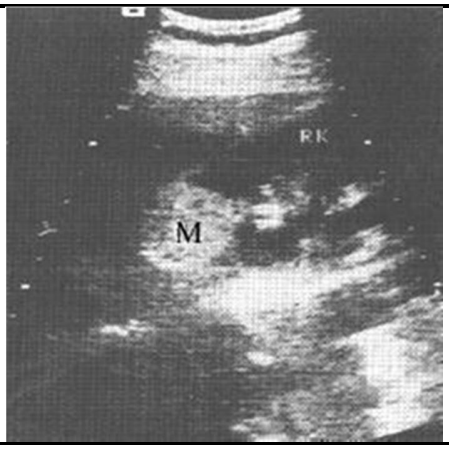


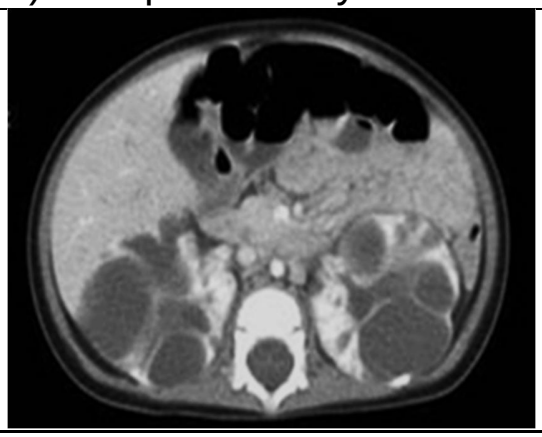
F- Renal & Retroperitoneal Involvement

1) Renal Angiomyolipoma:

- composed of fat, smooth muscles and aggregates of blood vessels.
- Usually large, multiple and bilateral
- US: Echogenic focal lesion
- CT: noncalcified cortical tumors containing fat
- Complication: Pseudo-aneurysms, intrarenal, perinephric, or retroperitoneal hemorrhage

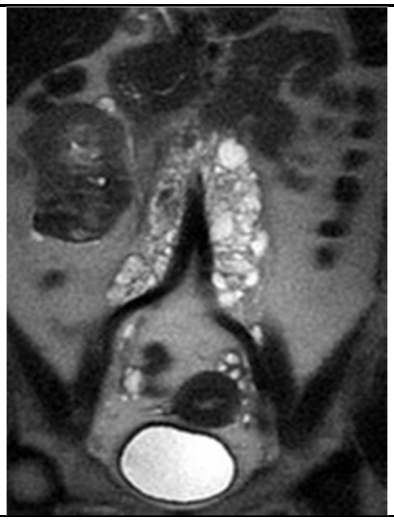




2) Multiple Renal Cysts:	3) Renal cell carcinoma:
	

4) Retroperitoneal LAM

- Thick- or thin-walled cystic lesions can be found in the retroperitoneum, which may reflect dilatation of lymph vessels due to obstruction

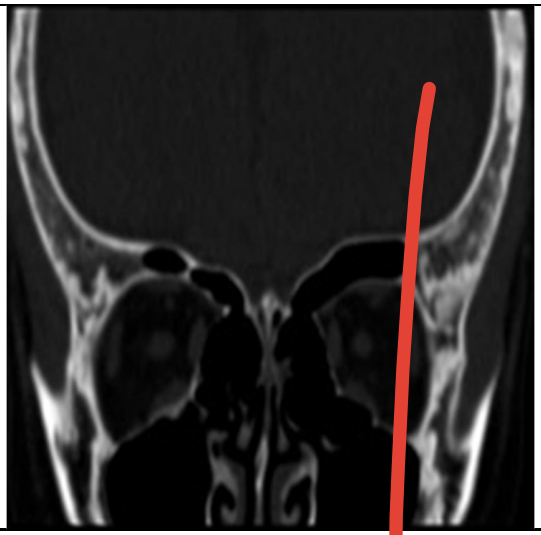


G- Hepatobiliary Involvement

- Hepatomegaly
- AMLs
- Lipomas
- Hamartomas
- Fibromas.

H- Skeletal Involvement

- Cyst like lesions
- Hyperostosis of the inner table of the calvaria
- Osteoblastic changes
- Periosteal new bone formation
- Scoliosis.



D- Sturge-Weber Syndrome

- Sturge-Weber syndrome, also known as encephalotrigeminal angiomatosis, is a rare neurocutaneous disorder.

- The pathogenesis is believed to be related to a vascular steal phenomenon secondary to an extensive cortical pial angiomatous malformation

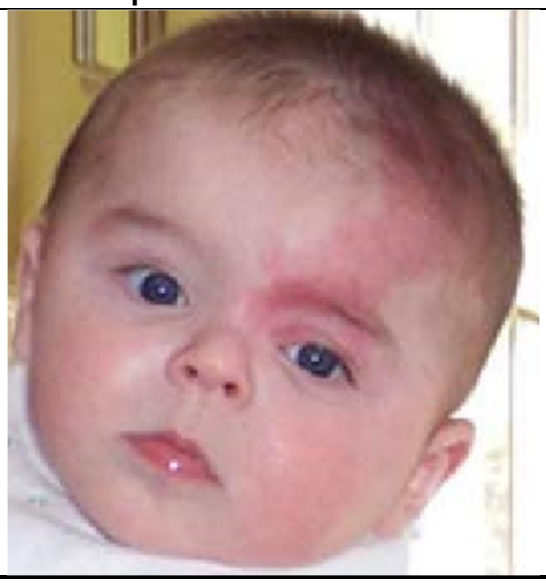
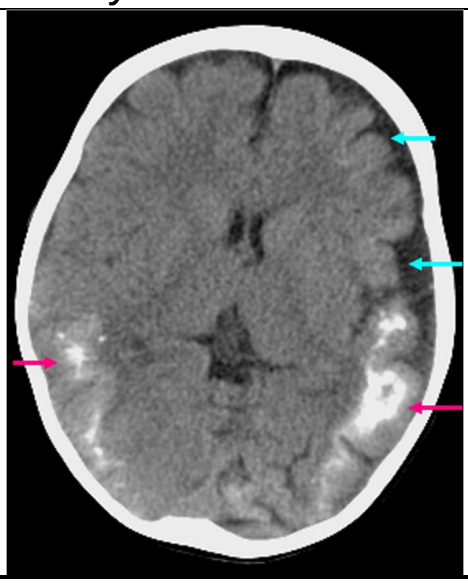
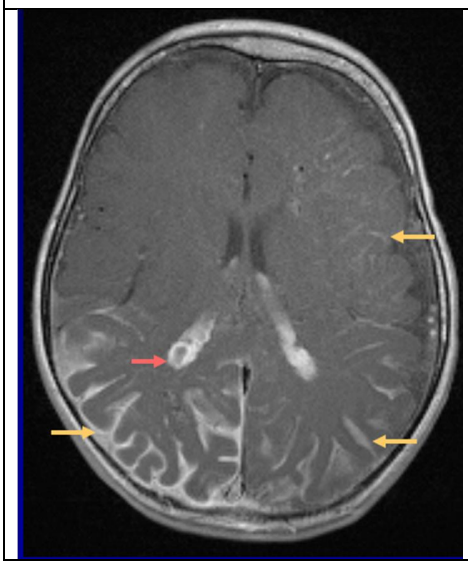
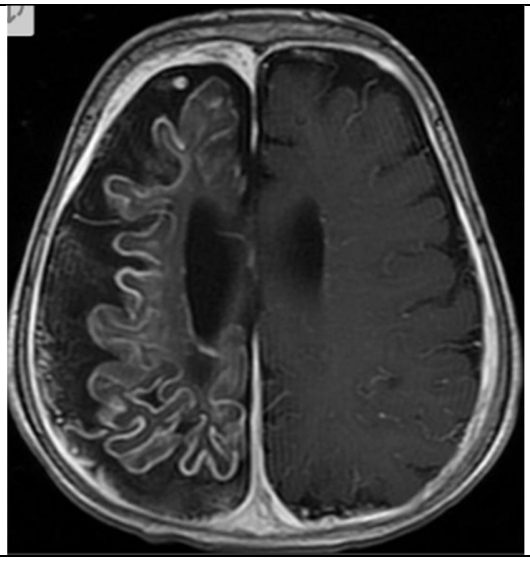
Clinical picture:

- Facial cutaneous hemangioma (port wine stain) along the distribution of the ophthalmic division of the trigeminal distribution.
- Epilepsy
- Mental retardation

SWS Type	Clinical Features
1 ("classic")	Facial and intracranial manifestations
2	Facial lesion only (primarily dermatological)
3	Intracranial manifestations *without* facial

Imaging findings:

- CT Bone window / T2*:
  - Dense gyriform calcifications, which more commonly affect the parieto-occipital cortical areas or the choroid plexus
  - Tram-track sign of cortical and subcortical calcification
  - Thickening of the calvaria as an indirect feature of loss of the brain substance
- T1+C:
  - Leptomeningeal enhancement + Gyriform enhancement; reflecting pial angiomatosis.
  - Enlargement and abnormally avid enhancement of the ipsilateral choroid plexus.
- T2/ FLAIR:
  - Areas of gliosis likely related to chronic ischemia.
  - Cerebral ipsilateral hemiatrophy as a consequence of vascular steal phenomena of the pial angioma on the surrounding cortical structures.

Facial port-wine stain	CT: Gyriform Calcification
	
Axial T1+C: Choroid plexus enlargement + leptomeningeal enhancement	Late stage: Cerebral hemiatrophy
	

E- Von Hippel-Lindau syndrome (VHL)

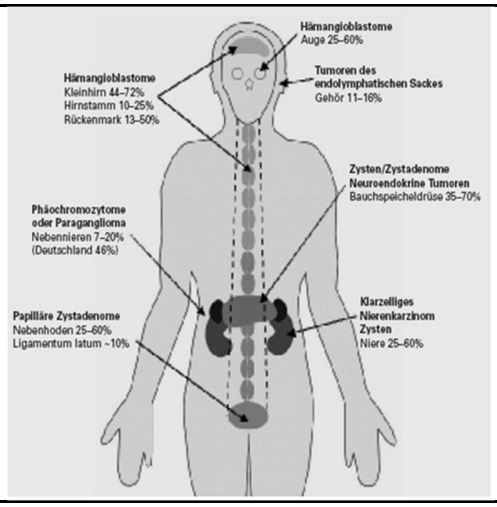
- Rare, AD, inherited, neurocutaneous dysplasia
- The condition is associated with inactivation of a tumor suppression gene located on chromosome 3p25.5
- Sex: M=F
- Age: Present after 2nd decade.

Diagnostic Criteria For VHL

A) > 1 CNS hemangioblastoma.
(B) 1 CNS hemangioblastoma + Visceral manifestations of VHL disease.
(c) Any manifestation + a known family history of VHL disease.

Most common causes of death:

- Renal cell carcinoma.
- Neurologic complications from cerebellar hemangioblastoma.



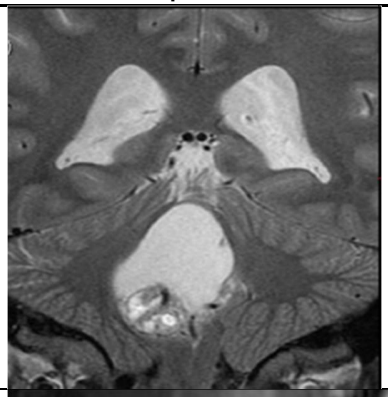
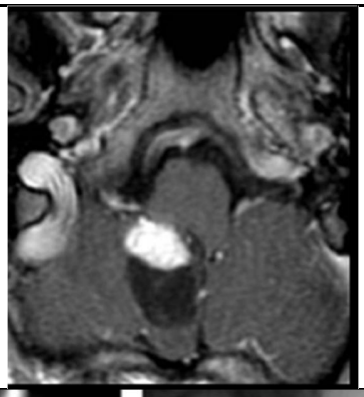
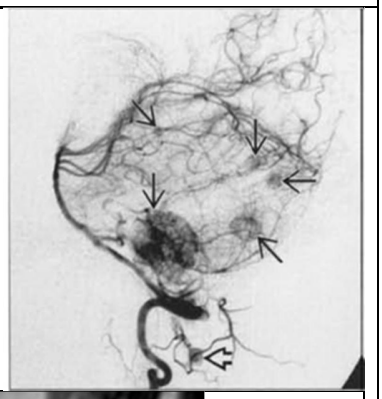
Manifestations of VHL Disease according to Prevalence

Manifestations	Prevalence (%)
Pancreatic cysts	50–91
Cerebellar hemangioblastoma	44–72
Renal cysts	59–63
Retinal hemangioblastoma	45–59
Renal cell carcinoma	24–45
Spinal cord hemangioblastoma	13–59
Pheochromocytoma	0–60
Neuroendocrine tumor of the pancreas	5–17
Serous cystadenoma of the pancreas	12
Medullary hemangioblastoma	5
Papillary cystadenoma of the epididymis	10–60*

*Of male patients.

1) CNS Hemangioblastoma

- is one of the most common manifestations of VHL disease.
- Typical sites are:
  - Cerebellum (44%–72%)
  - Spinal cord (13%–59%)
  - Medulla (5%)
  - Supratentorial lesions are less common

2) Retinal Hemangioblastomas

- The prevalence = 45% - 59%
- 50% cases being bilateral

3) Endolymphatic Sac Tumors

CT:

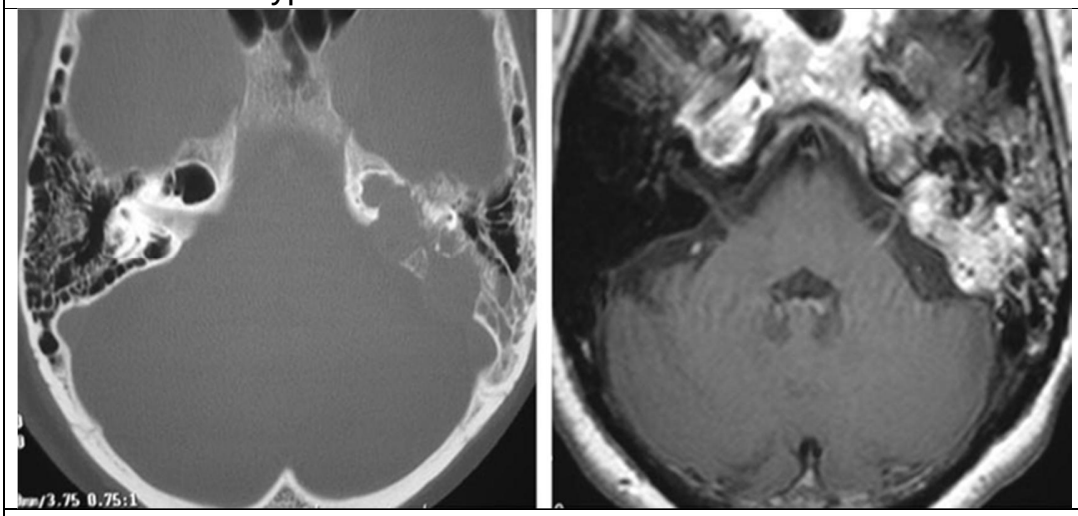
- The bone destruction is geographic or moth-eaten and the intratumoral bone is stippled, reticular, or spiculated
- There is a peripheral rim of calcification.

MRI:

- The signal intensity is heterogeneous with either a peripheral rim of high signal intensity or hyperintense foci.
- Contrast enhancement is heterogeneous.

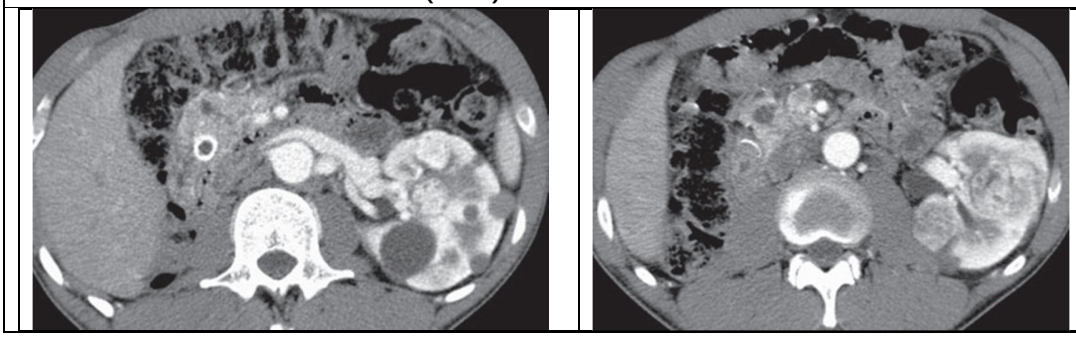
Angiogram:

- Reveal a hypervascular tumor.



4) Renal Cysts and Tumors

- Renal cysts occur in 59%–63% of patients
- Renal cell carcinoma (RCC) in 24%–45%.

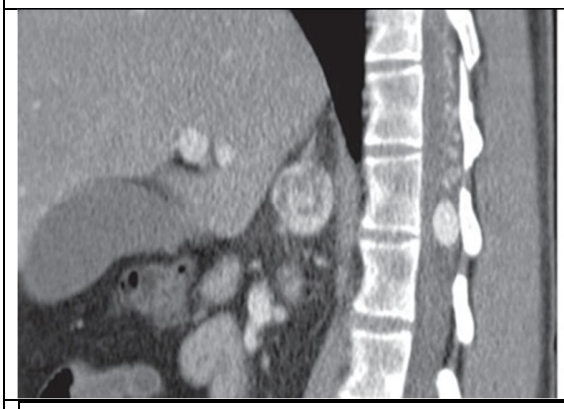


5) Pancreatic Cysts and Tumors

- Pancreatic cysts (50%–91%)
- Serous microcystic adenomas (12%)
- Neuroendocrine tumors (islet cell tumors) 5%–17%
- Rarely adenocarcinomas

6) Pheochromocytoma

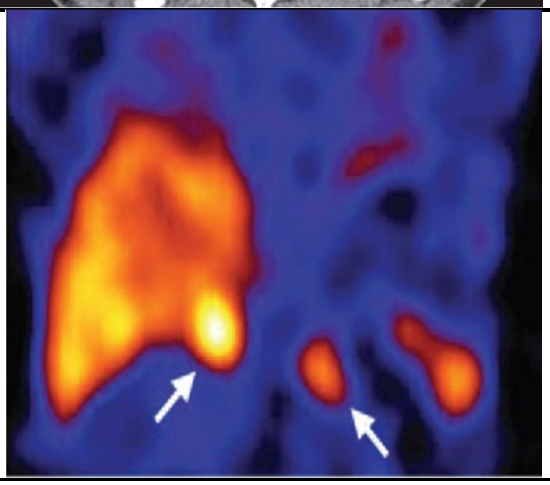
- Pheochromocytomas arise from the neural crest and produce elevated levels of catecholamines in the serum and urine.



131I MIBG scanning :

75%– 95% sensitive

98%–100% specific



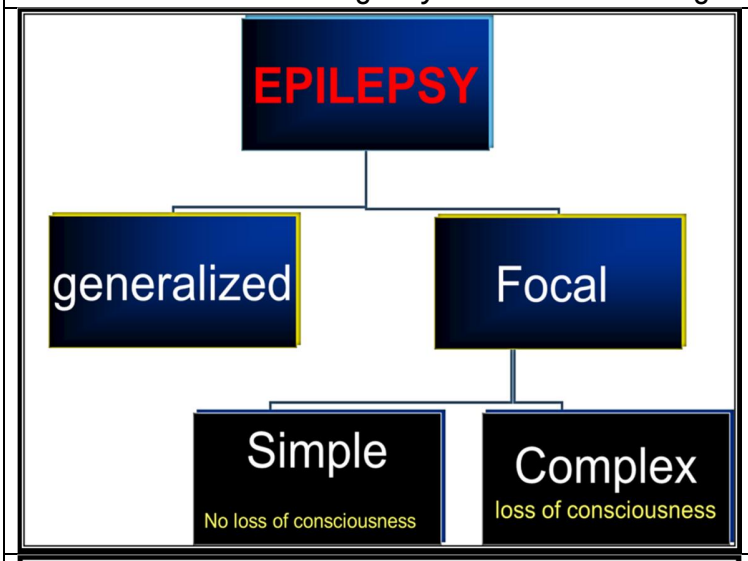
7) Papillary Cystadenomas Of The Epididymis

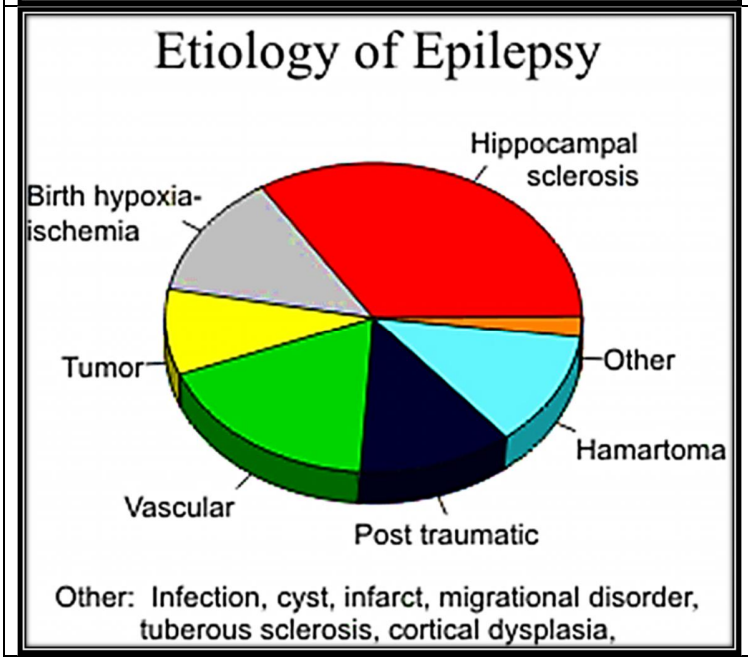
- is most commonly located in the head of the epididymis but may also involve the spermatic cord.

Epilepsy

Aim Of Neuroimaging In Epilepsy Patients:

- Identify underlying structural abnormalities that require specific treatment (ie, surgery in most instances)
- To aid in formulating a syndromic or etiologic diagnosis.





❖ Temporal Lobe Epilepsy (TLE)

- is the most common type of partial epilepsy
- It is divided into 2 broad groups:

Medial temporal epilepsy (most common)

- involves the mesial temporal lobe structures
- most frequently due to mesial temporal sclerosis

Lateral temporal epilepsy can be caused by:

- Tumors: (Ganglioglioma, Gangliocytoma, Dysembryoplastic neuroepithelial tumor (DNET))
- Vascular malformations
- Cortical dysplasias

A- Mesial Temporal Sclerosis (MTS)

- MTS is one of the most common causes of epilepsy in the adolescent and young adult population.

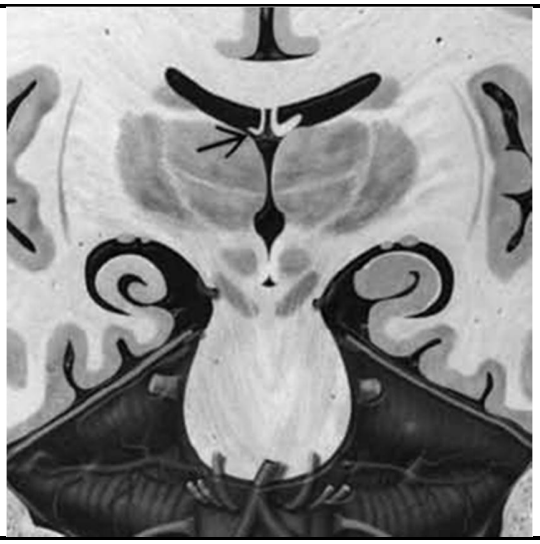
MTS Characterized by:

Neuronal loss + Gliosis + Atrophy:

Hippocampus

± Ipsilateral Fornix

± Mamillary Body



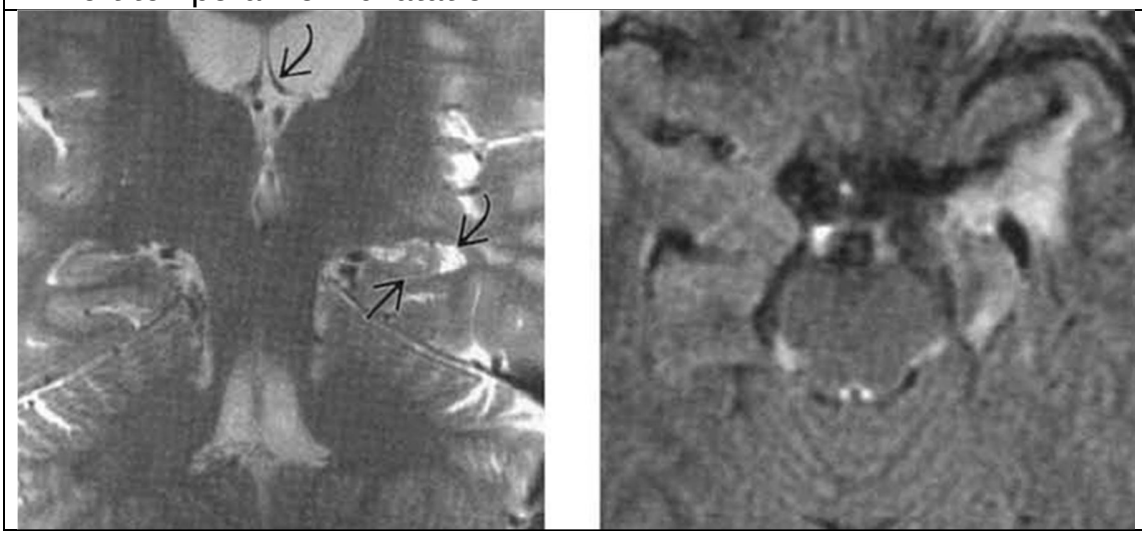
Imaging findings:

- Hippocampus T2/FLAIR hyperintense signal
- Hippocampus atrophy
- Loss of gray-white differentiation/internal architecture in hippocampus
- Bilateral in 20%

2ry Findings of MTS:

- Temporal lobe volume loss
- Dilatation of the temporal horn
- Narrowed collateral white matter
- ± Small fornix
- Mamillary body atrophy

- left hippocampus atrophy and abnormal T2/FLAIR hyperintensity
- Left Fornix atrophy
- Left temporal horn dilatation



B- Temporal Lobe Tumors (see chapter of tumors)

- Ganglioglioma
- Pleomorphic Xanthoastrocytoma (PXA)
- Pilocytic Astrocytoma
- Dysembryoplastic Neuroepithelial Tumour (DNET)

❖ Neuronal Migration and Proliferation Anomalies (Discussed before)

Disorders of cortical formation according to stages

Stage	Cause	Disorder
Proliferative	Decreased proliferation	Microlissencephaly
	Increased proliferation	Hemimegalencephaly
	Abnormal proliferation	Focal cortical dysplasia
Migration	Undermigration	Complete (classic) lissencephaly
	Overmigration	Congenital muscular dystrophy
	Ectopic migration	Heterotopia
Organization	Deranged organization	Polymicrogyria
		Schizencephaly

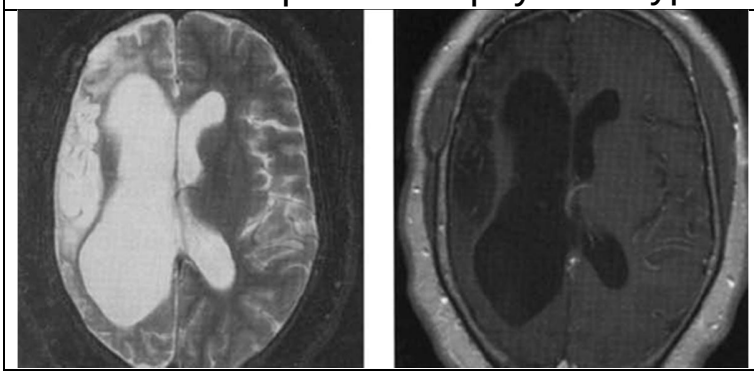
❖ Hypoxic ischemic Encephalopathy (Discussed before)

❖ Phacomatoses Neurocutaneous (Discussed before)

❖ Rasmussen Encephalitis

- Rare childhood syndrome
- Chronic progressive unilateral inflammation of the brain of uncertain cause.
- Characterized by medically refractory focal seizures and progressive multifocal neurologic symptoms such as hemiplegia and cognitive impairment.

Cerebral Hemisphere Atrophy + T2 Hyperintensity



❖ Limbic Encephalitis

- Limbic encephalitis manifests clinically as limbic system dysfunction with subacute onset of psychiatric symptoms, seizures, and memory loss

- Limbic encephalitis had been traditionally described in ***adults***, in whom it was initially thought to be ***paraneoplastic syndrome*** associated with carcinomas

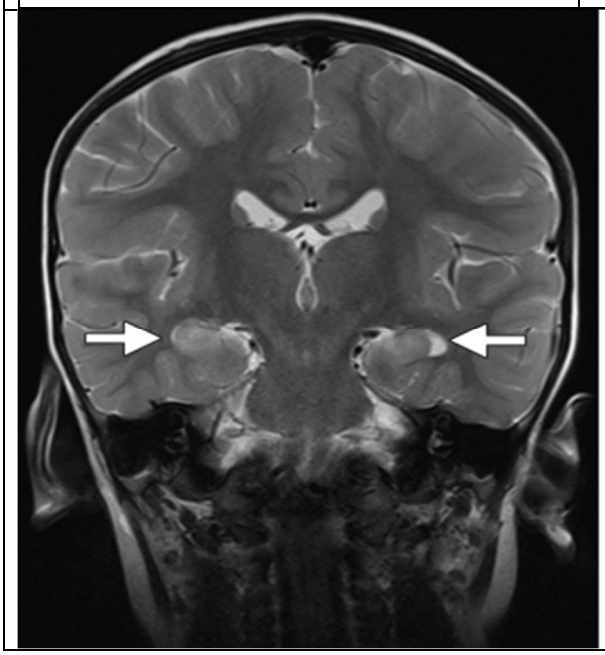
Acute stage:

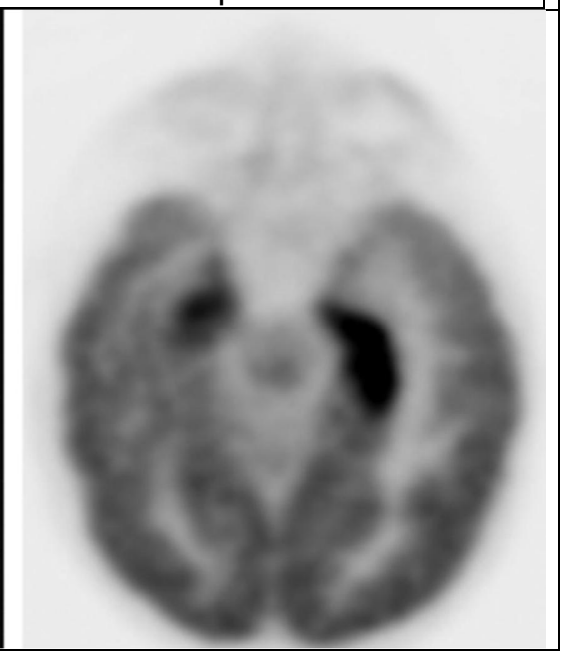
- T2-hyperintense mesial temporal lesions.
- PET: marked corresponding hypermetabolism

Later stage:

- T2 decreased hippocampal signal intensity + volume loss
- FDG decreased uptake in corresponding regions

Coronal T2: bilateral areas of hyperintensity in hippocampus (arrows).	FDG PET image shows corresponding appreciable increased metabolism bilaterally in the mesial temporal lobes.





❖ STATUS EPILEPTICUS

- Status epilepticus is defined as continuous seizure activity lasts > 30 min.

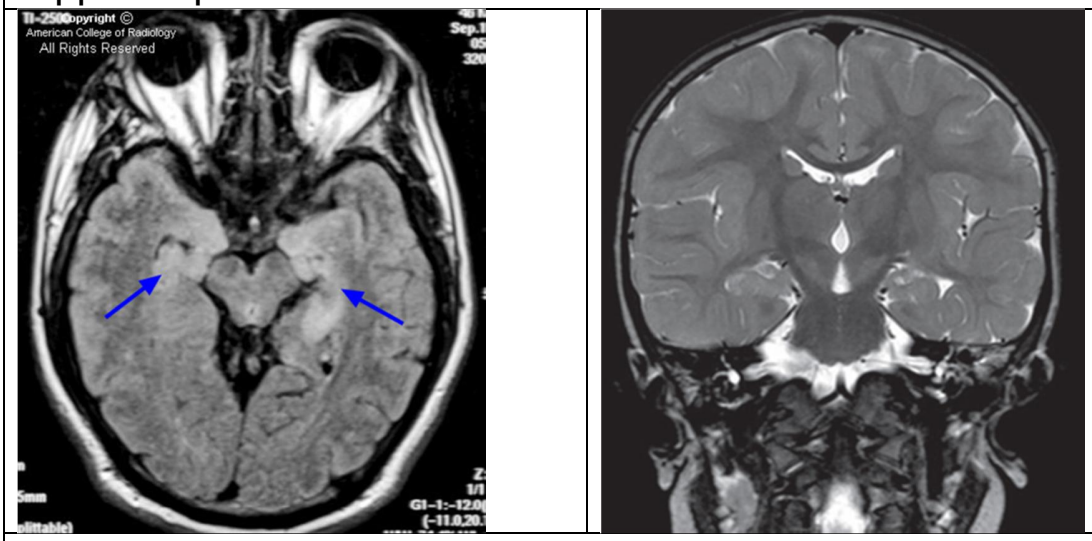
Location:

- Typically cortex and/or subcortical white matter
- Hippocampus
- Splenium of the corpus callosum

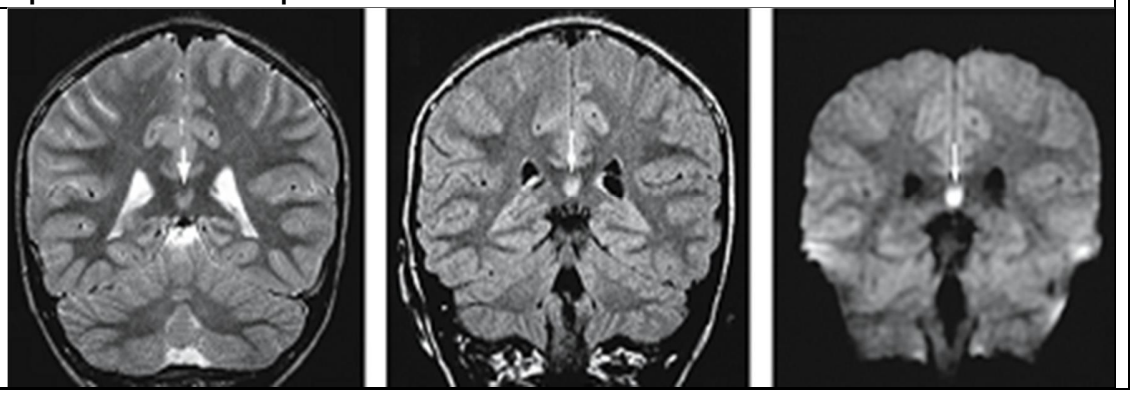
Imaging findings:

- T2: hyperintensity in (GM) and/or subcortical (WM)
- Mild mass effect
- DWI: Restricted diffusion
- ADC: Decrease

Hippocampus



Splenium of corpus callosum

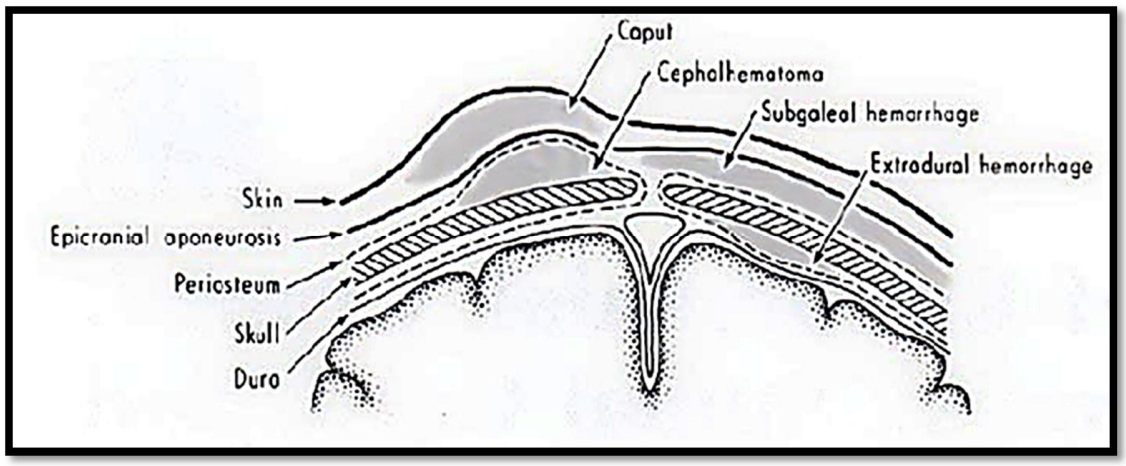


Head Trauma

I- 1ry signs => Centripetal approach (outside to inside)
A) Extracranial Haematoma
B) Calvarial Fractures
C) Extra-axial Hemorrhage: 1) Epidural hematoma 2) Subdural hemorrhage 3) Subdural hygroma 4) Subarachnoid hemorrhage
D) Intra-axial Injury: 1) Cortical contusion 2) Diffuse axonal injury 3) Intaraparenchymal hematoma 4) Intraventricular hemorrhage
II- 2ry Signs
A. Cerebral herniation B. Diffuse cerebral edema C. Post-traumatic infarction D. Post-traumatic hydrocephalus
III- Vascular Complications
A. Intimal irregularity B. Intramural hematoma C. Dissection D. Pseudoaneurysm E. Occlusion F. Transection with extravasation

I- 1ry Signs of Head Trauma

A) Extracranial Hematoma:



i) Caput Succedaneum:

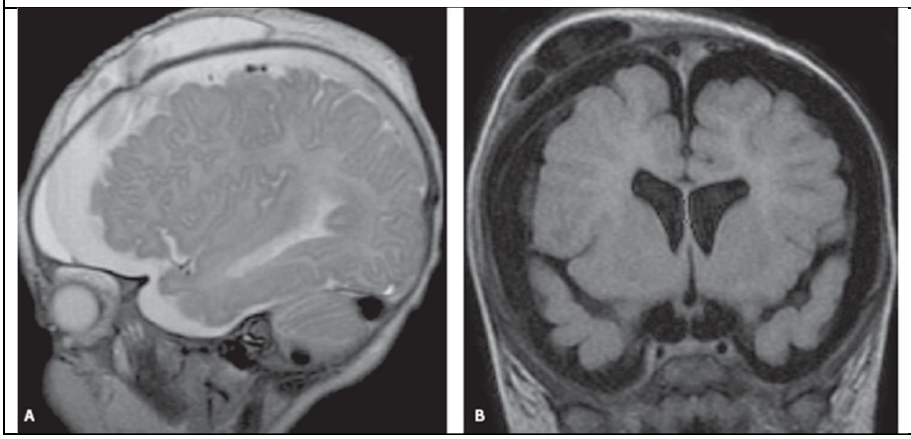
- Within the skin
- Spontaneously resolved
- Cross the sutures

ii) Subgaleal haemorrhage

- The subgaleal space extends from the orbital ridges to the nape of the neck and laterally to the ears.
- Beneath the apenoureses
- Can cross the sutures.

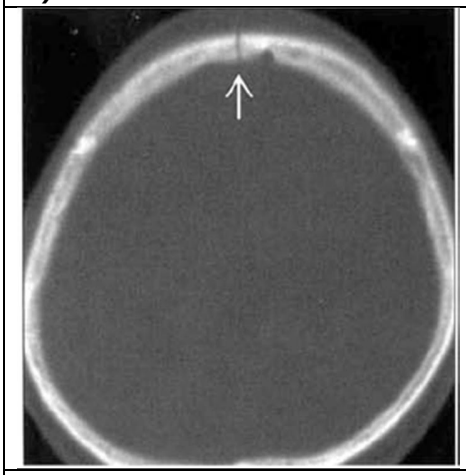
iii) Cephalhematoma:

- Subperiosteal
- Not cross the sutures

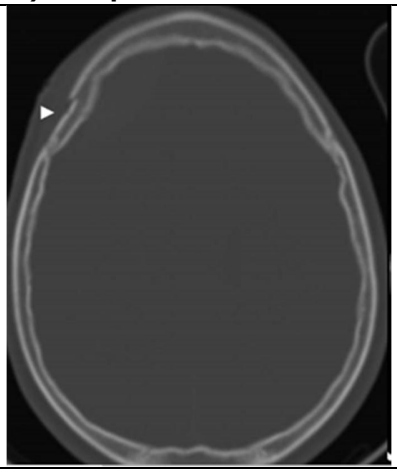


B) Fractures:

1) Fissure fracture

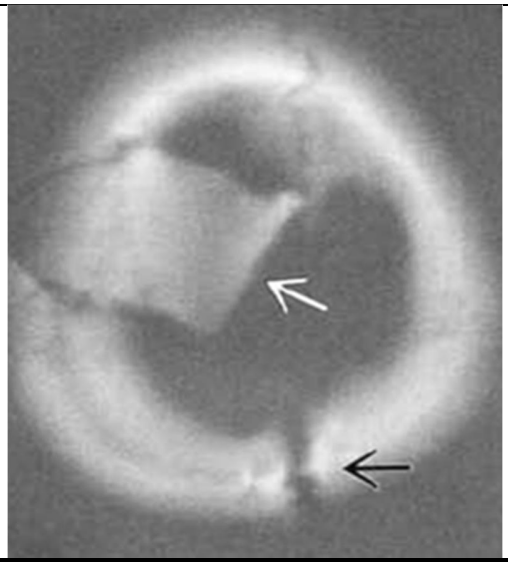


2) Depressed Fracture



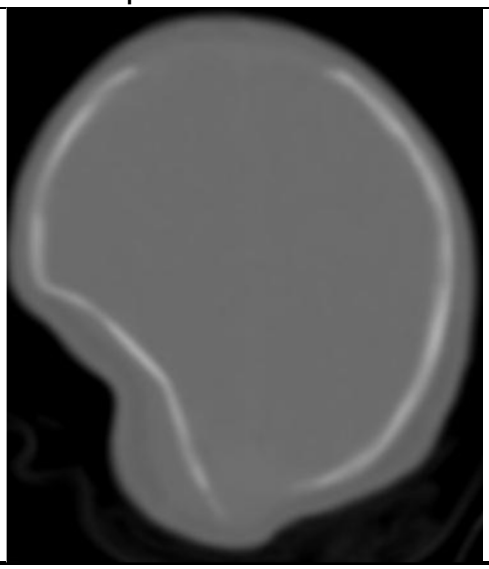
3) Diastatic Fracture

Wide sutures > 4mm

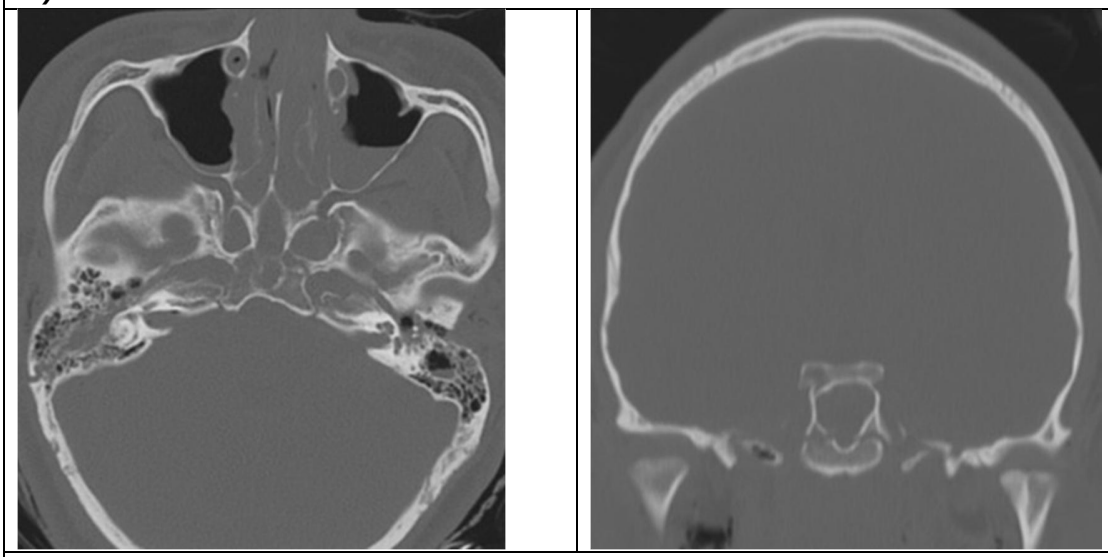


4) Ping-pong Fracture

Depressed fracture of the infant skull caused by inner buckling of the calvarim
+ intact periosteum and dura

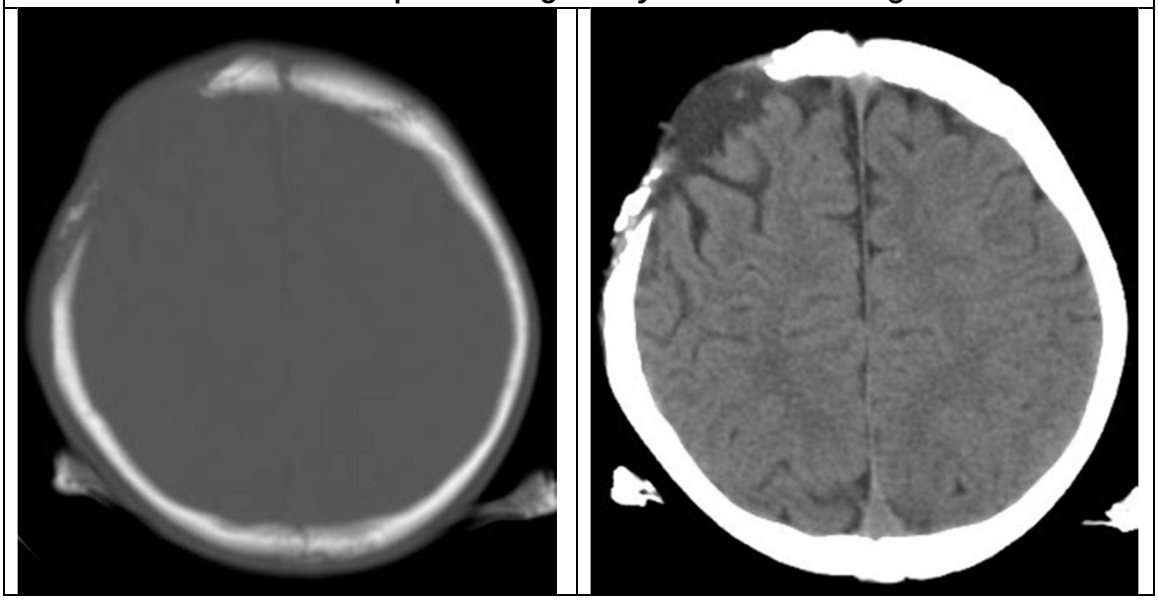


5) Skull Base Fracture



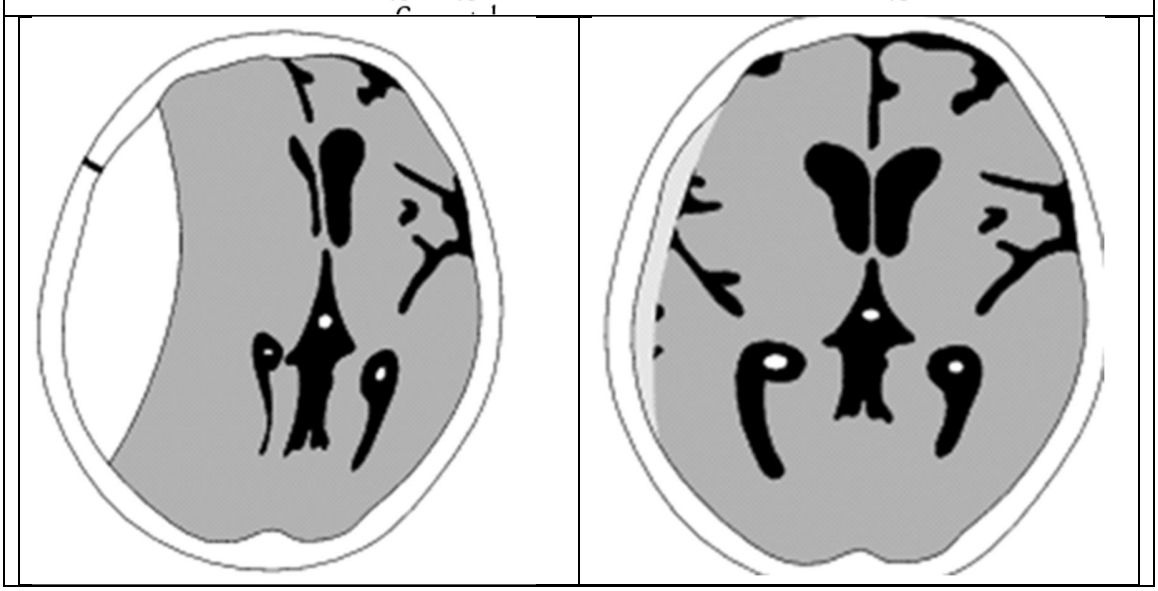
6) Growing Fracture

Calvarial fracture + Leptomenigeal cyst => widening the fracture



C) Extra-Axial Hemorrhage

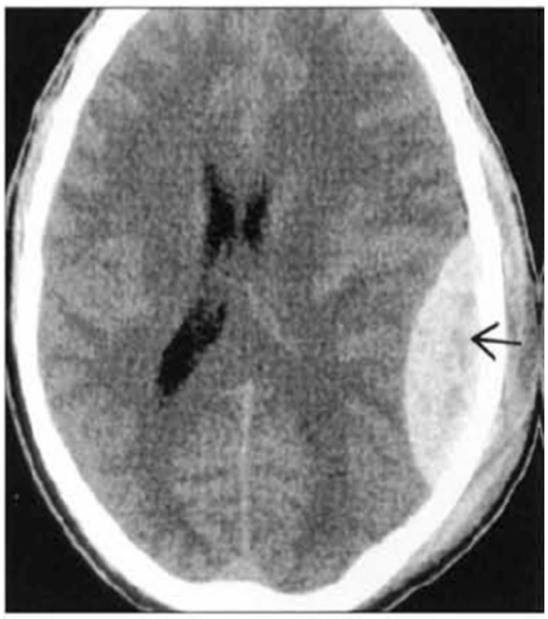
	EPIDURAL HAEMATOMA	SUBDURAL HAEMATOMA
Incidence	1-4% of trauma cases; 10% of fatal trauma cases	10-20% of all trauma cases; 30% of fatal trauma cases
Aetiology	Associated fractures in 85-95% of cases; Laceration of middle meningeal artery/dural venous sinus in 70-80% of cases.	Tearing of the cortical veins of the pons
Site	Between skull and dura mater; Crosses the dura mater but not the cranial sutures; 95% are supratentorial 5% are subtentorial; 5% bilateral	Between dura mater and arachnoid mater; Crosses the cranial sutures but not the dura mater; 95% are supratentorial; 5% are bilateral
CT findings	Biconvex (lens) shape; Shifts the white-grey matter interface; 66% are hyperdense; 33% are mixed (hyper-/hypodense)	*Acute*: 60% hyperdense; 40% mixed (hyper-/hypodense) *Subacute*: isodense; *Chronic*: hypodense



1) Extradural Haematoma

- Commonly (95%):
  - Unilateral / Coup site = Impaction site
  - Tempro-parietal fracture
  - Middle Meningeal Artery bleeding.
  - Tempro-parietal region

- Biconvex = lenticular
- Acute = Hyperdense
- "Swirl sign" = actively/rapidly bleeding hematoma
  - Mixed high and low attenuation blood within the hematoma
  - Low attenuation blood is hyperacute unclotted blood
  - High attenuation blood is already clotted
- Don't cross sutures
- Can cross falx & tentorium



2) Subdural Haematoma

- Result from tearing of cerebral veins

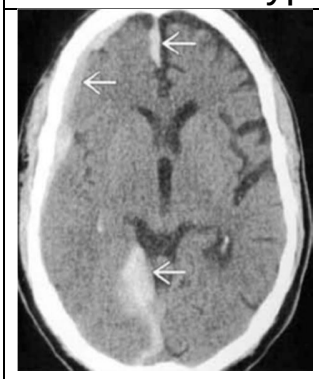
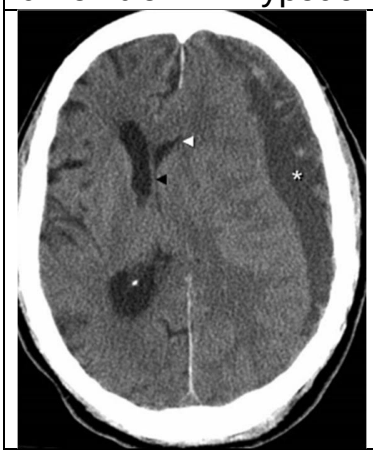
- crescentic extra-axial collection
- May cross sutures, not dural attachments
- May extend along falx & tentorium

CT density of the blood:

- Acute: denser than the brain.
- Subacute: isodense to the brain.
- Chronic: less dense than brain.

Indirect signs of subacute subdural hematoma:

- Midline shift.
- Compression of the ipsilateral ventricle.
- Contralateral ventricular enlargement.
- Effacement of cerebral sulci.
- Medial displacement of the junction between white and grey matter.

Acute SDH = Hyperdense	Subacute SDH = Isodense
	
Chronic SDH = Hypodense	SDH of different ages = Hematocrit level
	

3) Subarachnoid Hemorrhage (SAH)

Causes of SAH:

i. Trauma (most common cause of SAH)

ii. Non traumatic SAH = Spontaneous SAH:

- Ruptured aneurysm 85% (most common cause of non-traumatic (SAH)
- Non-aneurysmal (perimesencepalic) haemorrhage - 10%
- Vascular malformations
- Pituitary apoplexy
- Cocaine use

Complications of SAH:

i) Acute hydrocephalus

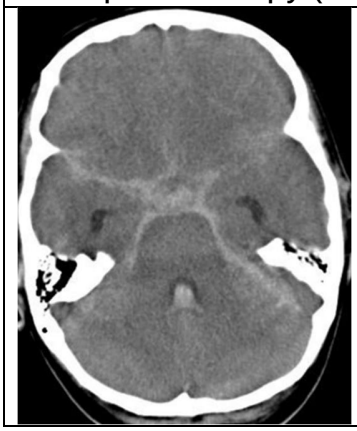
- Rare
- usually obstruction of aqueduct or 4th ventricular outlet by clotted SAH
- Obstructive, non-communicating hydrocephalus
- Asymmetric ventricular dilatation

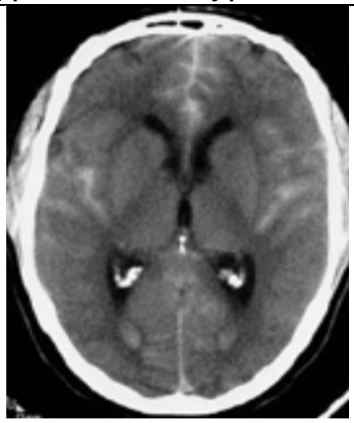
ii) Delayed hydrocephalus

- Arachnoid granulation defect in CSF resorption
- Obstructive, communicating hydrocephalus
- Symmetric ventricular dilatation

iii) Vasospasm

- May develop quickly (2-3 days post-injury)
- Peaks 7-10 days after injury
- Threat remains up to 2 weeks
- Uncommon cause of post traumatic infarct.
- Triple H therapy (Haemodilution, Hypertension, Hypervolaemia)





4) Subdural Hygroma

- Cerebrospinal fluid (CSF) accumulation in the subdural space

Causes:

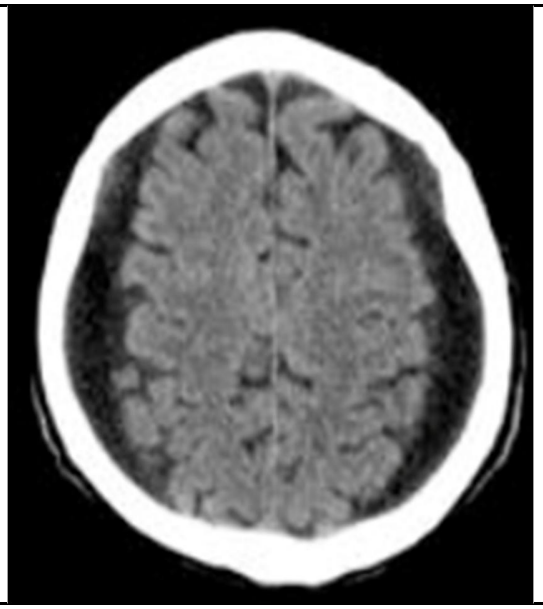
- Idiopathic: in pediatric patients
- Trauma: secondary to tear in the arachnoid mater
- Post-surgical, e.g. hematoma evacuation, ventricular drainage
- Spontaneous intracranial hypotension

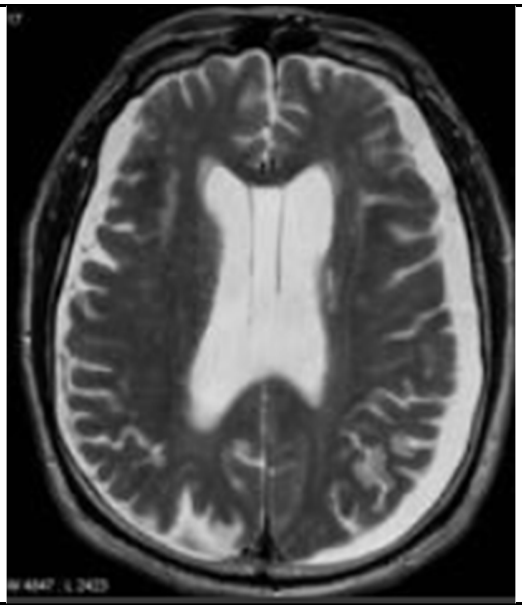
Location:

- Supratentorial cerebral convexities

Imaging findings:

- Crescentic CSF density/signal in subdural space
- Does not extend into the sulci.





5) Pneumocephalus

Air seen within the cranium:

- Subdural: sharply defined, lying superficially, adjacent to the midline.
- Subarachnoid: diffuse air bubbles.
- Intraventricular air is rarely seen.

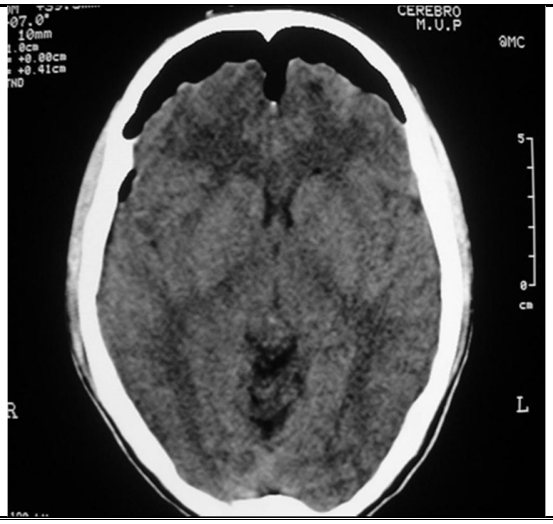
<u>Causes:</u>

- Commonly after the evacuation of a subdural hematoma.
- Skull base surgery
- Paranasal sinus surgery
- Posterior fossa surgery
- Head trauma

6) Tension Pneumocephalus

- Neurosurgical emergency.

- Mount Fuji sign on CT
- Is presence of air between the frontal tips separated by adhering cortical veins
- It suggests that the pressure of the air is at least greater than that of the surface tension of cerebrospinal fluid between the frontal lobes.



D) Intra-Axial Injury:

1) Cerebral Contusion

- Caused by traumatic contact of the cortical surface of the brain against the rough inner table of the skull.

Location:

- 50% temporal lobes
- 33% frontal lobe
- 25% parasagittal = "gliding" contusions
- Coup or a contrecoup location

Acute contusion:

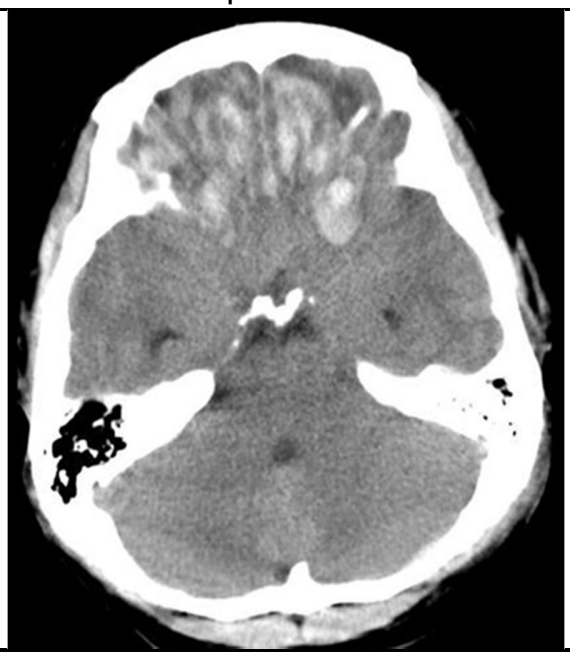
- Ill-defined, patchy or multifocal areas of petechial hemorrhage surrounded by cerebral oedema.

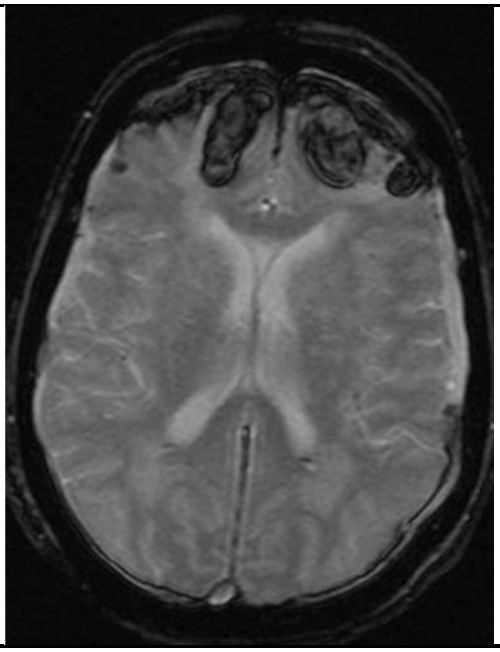
Subacute contusion:

- Ring enhancement

Chronic contusion:

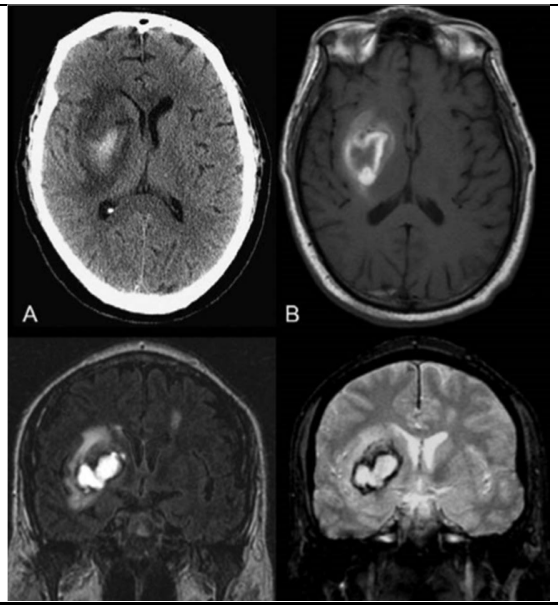
- Area of encephalomalacia





2) Intracerebral Hematoma

- A focal, well-defined, area of abnormal high CT density in the brain.



3) Diffuse Axonal Injury (DAI)

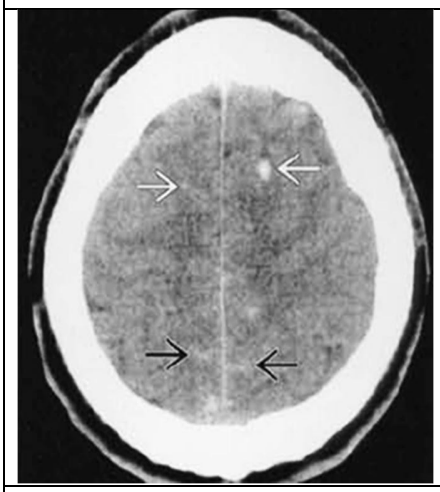
- Caused by rotational deceleration and subsequent reacceleration force that exceeds the limited elastic capacity of the axons.

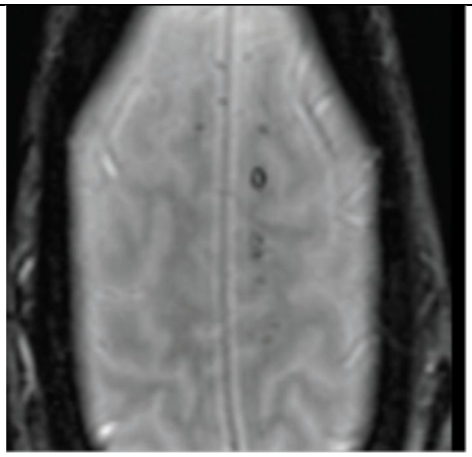
- Grade I DAI involves only the gray–white matter junctions.
- Grade II DAI involves the corpus callosum.
- Grade III (most severe) DAI involves the dorsolateral midbrain.

- CT is relatively insensitive for detection of DAI.
- Hemorrhagic DAI: Tiny hyperdense foci
- Nonhemorrhagic DAI: Tiny hypodense foci

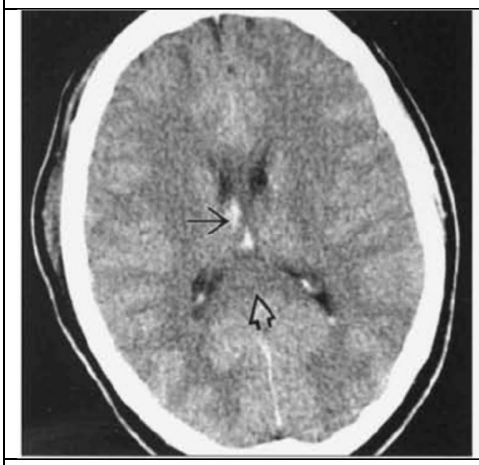
- MRI is much more sensitive to detect DAI:
- GRE is extremely sensitive for hemorrhagic DAI
- FLAIR is most sensitive for nonhemorrhagic DAI.
- DWI show restricted diffusion in acute DAI due to cytotoxic edema and cell swelling.

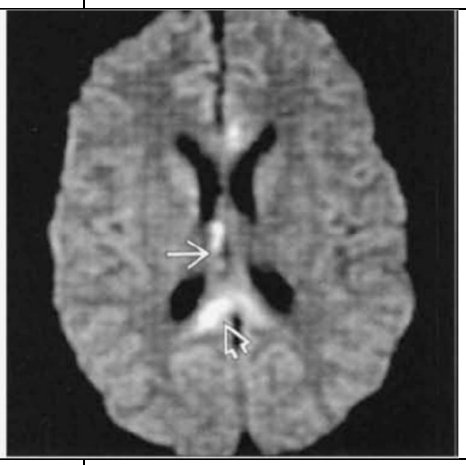
DAI Grade I



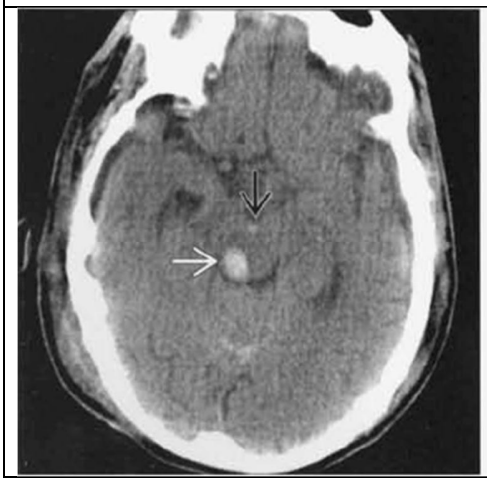


DAI Grade II





DAI Grade III



4) Intraventricular Hemorrhage:

- denotes the present of blood within the ventricular system of the brain.
- High morbidity due to the development of obstructive hydrocephalus.

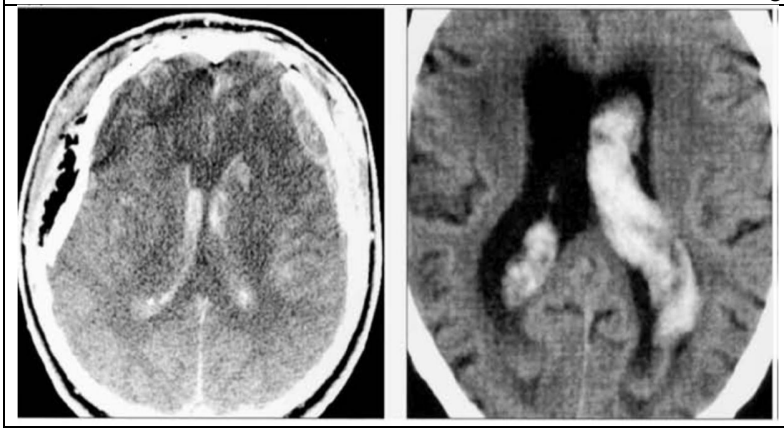
Causes:

Primary causes of intraventricular haemorrhage:

- Trauma due to tearing of subependymal veins
- Tumors:
    - Intraventricular tumours
        - Ependymoma
        - Choroid plexus / intraventricular metastases
    - Adjacent parenchymal tumours (e.G. GBM)
- Vascular malformations
    - Aneurysm
    - Arteriovenous malformations (AVM)
    - Subependymal cavernous malformations

Secondary causes of intraventricular haemorrhage include:

- Direct extension from intracerebral haemorrhage
    - Hypertensive haemorrhage, especially basal ganglia haemorrhage (common)
    - Lobar haemorrhage
- Direct extension form subarachnoid haemorrhage



II- 2ry Signs of Head Trauma

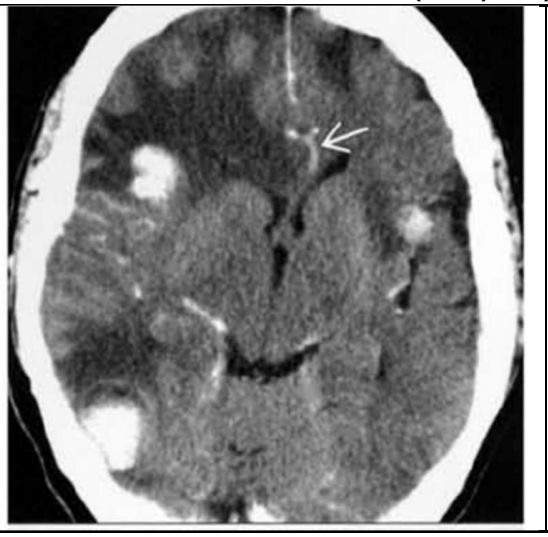
A) Cerebral Herniation

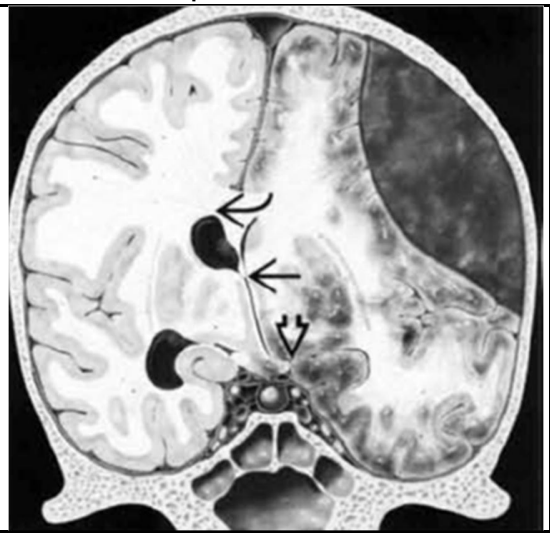
1) Subfalcine herniation
2) Unilateral descending transtentorial herniation (DTH)
3) Bilateral descending transtentorial herniation (central DTH)
4) Ascending transtentorial herniation
5) Tonsillar herniation
6) Transalar herniation
7) Transdural / transcranial herniation

1) Subfalcine Herniation:

- Secondary to unilateral frontal lobe mass effect
- Herniation of cingulate gyrus under falx

⇨ Ipsilateral lateral ventricle compressed
⇨ Foramen of monro obstructs
⇨ Contralateral lateral ventricle enlarges
⇨ Anterior cerebral arteries (ACA) displaced + Compression

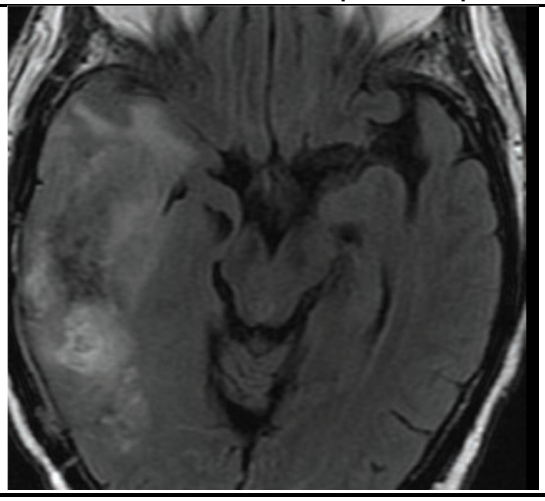


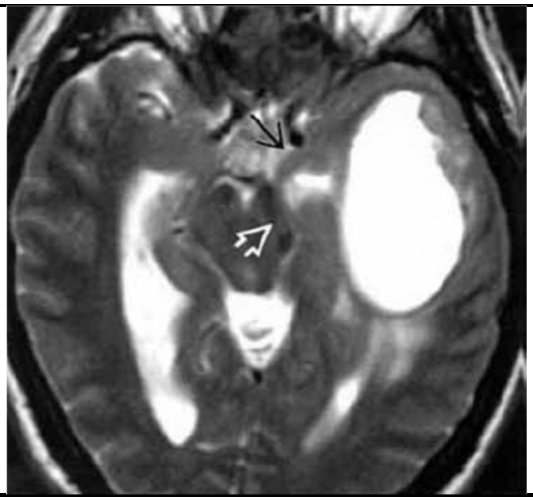


2) Unilateral Descending Transtentorial Herniation (DTH) = Uncal Herniation

- Herniation of medial temporal lobe inferiorly through incisura
- Secondary to unilateral mass effect on temporal lobe

⇨ Medial temporal lobe slides under tentorium
⇨ Uncus, parahippocampal gyrus displaced inferomedially
⇨ Ipsilateral suprasellar cistern effaced
⇨ Ipsilateral ambient, CPA cisterns enlarge
⇨ Ipsilateral CN III paresis
⇨ Compression of PCA
⇨ Upper brainstem Duret hemorrhages caused by shearing of perforating vessels due to downward force on the brainstem.
⇨ Compression of contralateral cerebral peduncle against Kernohan's notch causes a hemiparesis ipsilateral to the herniated side.

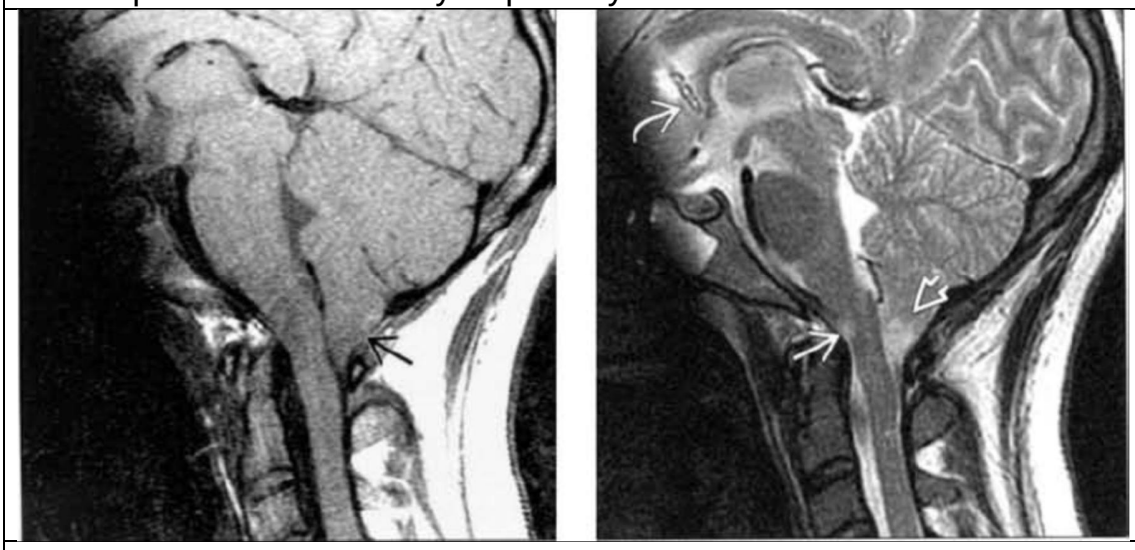




3) Cerebellar Tonsillar Herniation

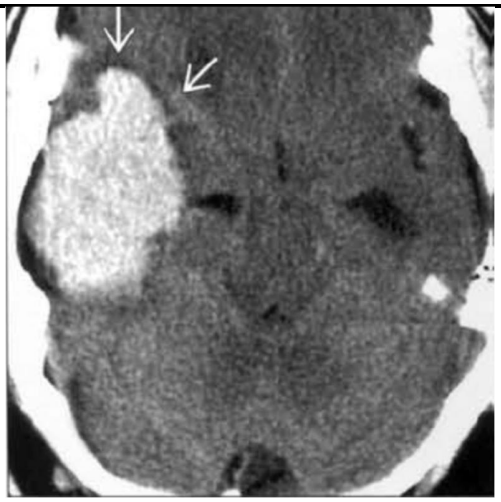
- Downward displacement of the cerebellar tonsils through foramen magnum
- "Peg_like" configuration of tonsils
- Displacement > 5 mm below foramen magnum

⇨ Compression of the medulla.
⇨ Compression of medullary respiratory centers is often fatal.



4) Transalar herniation

- Herniation across sphenoid wing
- Uncommon
- Few clinical symptoms
- Can be ascending (middle cranial fossa or temporal lobe mass) or descending (frontal lobe mass)
- Displacement of temporal & frontal lobes, middle cerebral artery across lesser sphenoid wing



B) Cerebral Edema

- Arise from increased cerebral blood volume (hyperemia), vasogenic edema, and increase in tissue fluid.
- Slit-like ventricles.
- Effacement of the basal cisterns and cortical sulci.

C) Post-traumatic Infarcts

- Downward trans-tentorial herniation => PCA occlusion
- Subfalcine herniation => ACA occlusion
- Central herniation => Basal perforating vessel occlusions

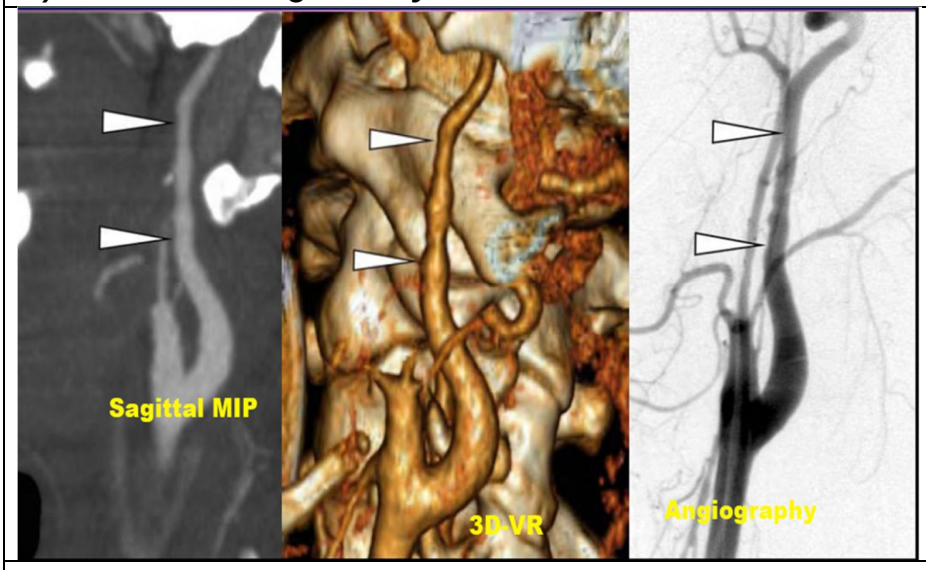
D) Post-traumatic Hydrocephalus:

- Occurs secondary to impaired CSF reabsorption by SAH at the level of:
  - Subarachnoid space
  - Aqueduct of sylvius
  - Outflow of 4th ventricle

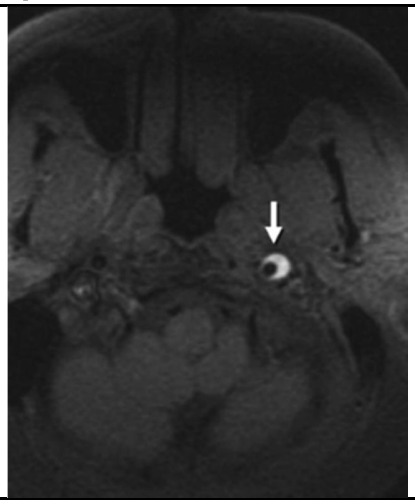
III- Vascular Complications of Head Trauma

A. Intimal irregularity
B. Intramural hematoma
C. Dissection
D. Pseudoaneurysm
E. Occlusion
F. Transection with extravasation

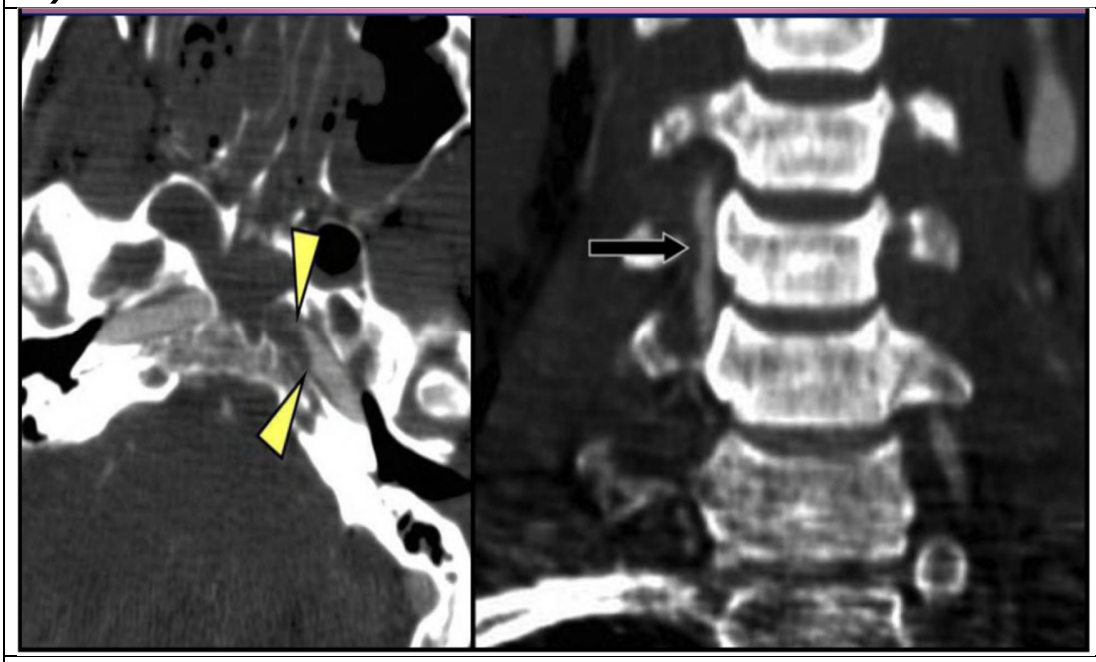
1) Intimal Irregularity



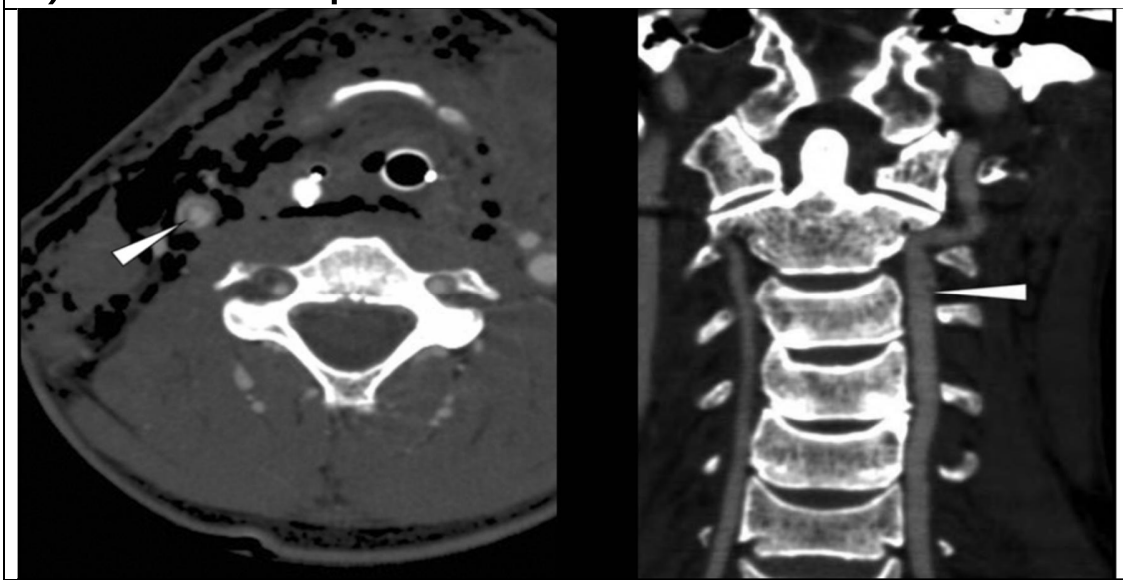
2) Dissection:



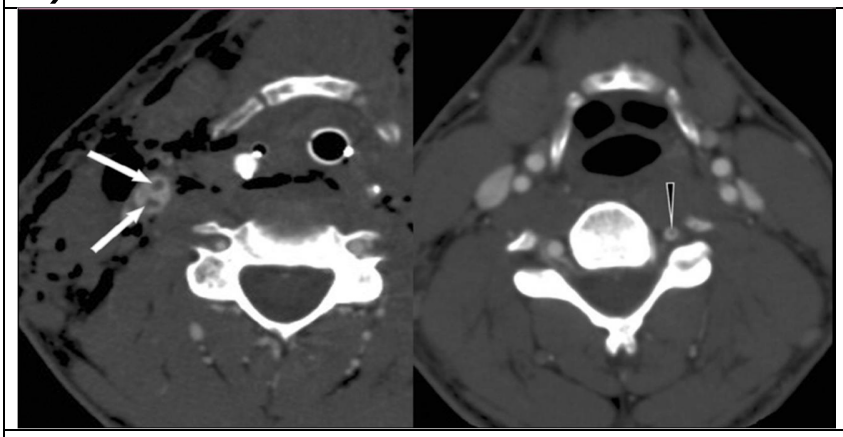
3) Intramural Hematoma



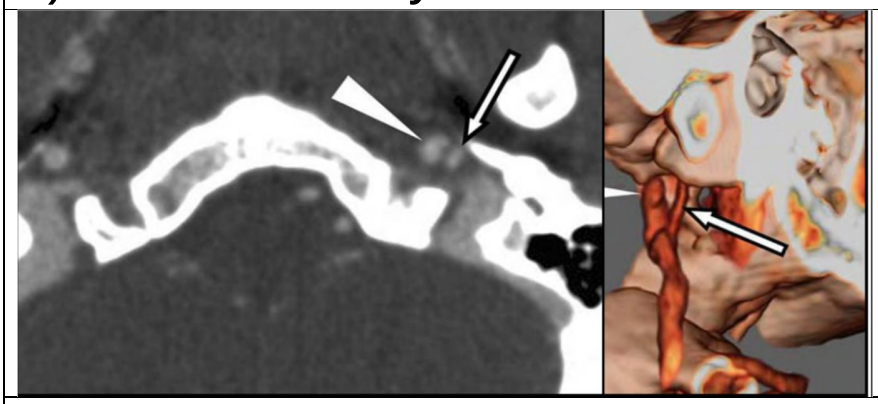
4) Intimal Flap:



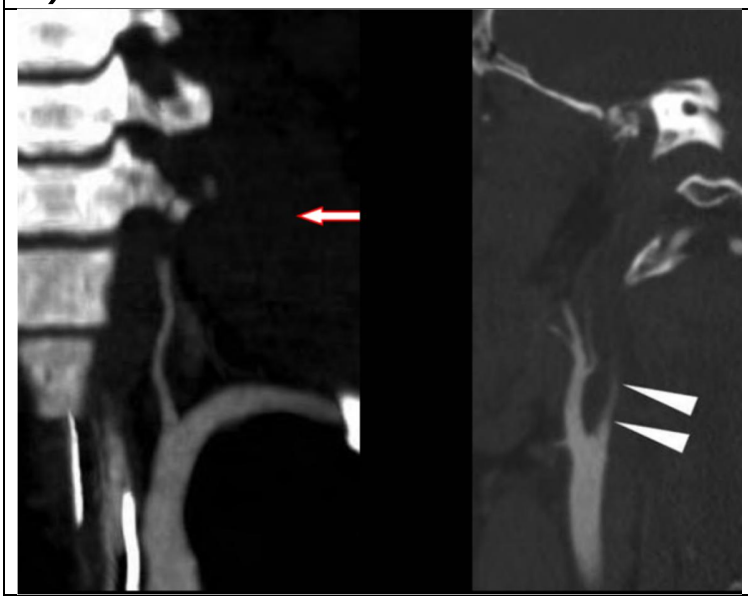
5) Intraluminal Thrombus



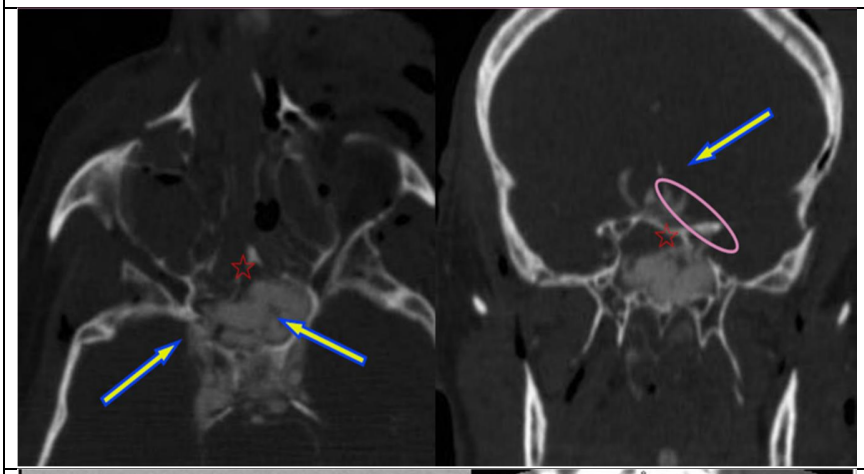
6) Pseudo-aneurysm

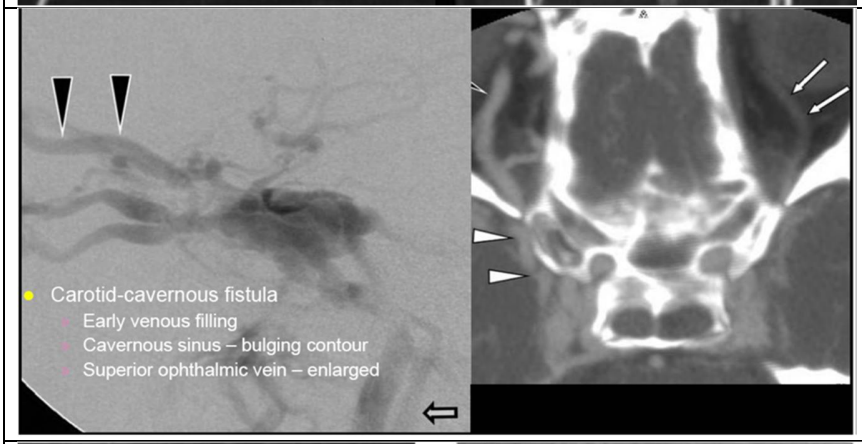


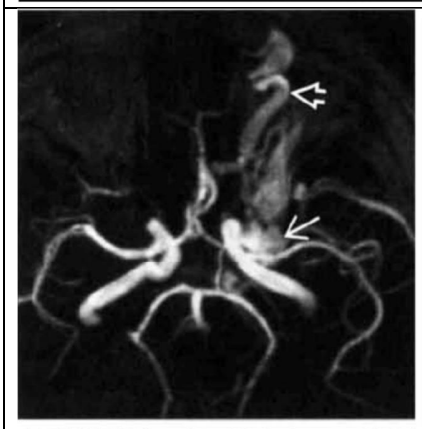
7) Occlusion



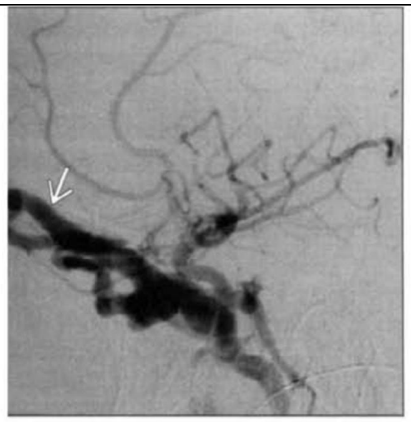
8) Transection
Carotid-Cavernous Fistula







Axial MIP MRA shows flow-related enhancement in left cavernous sinus (arrow) & flow in left superior ophthalmic vein (open arrow).



Lateral selective ICA conventional angiogram shows contrast immediately filling the cavernous sinus and draining via a very large superior ophthalmic vein (arrow).

Cerebrovascular Stroke

1) Ischemic Stroke

2) Intracerebral Hemorrhage

3) Subarachnoid Hemorrhage

4) Veno-occlusive Disease

I- Cerebrovascular Ischemic Stroke

- Stroke is a leading cause of death and disability worldwide.

❖ WHO definition of stroke

- "Rapidly developing clinical signs of focal (at times global) disturbance of cerebral function, lasting more than 24 h or leading to death with no apparent cause other than that of vascular origin"

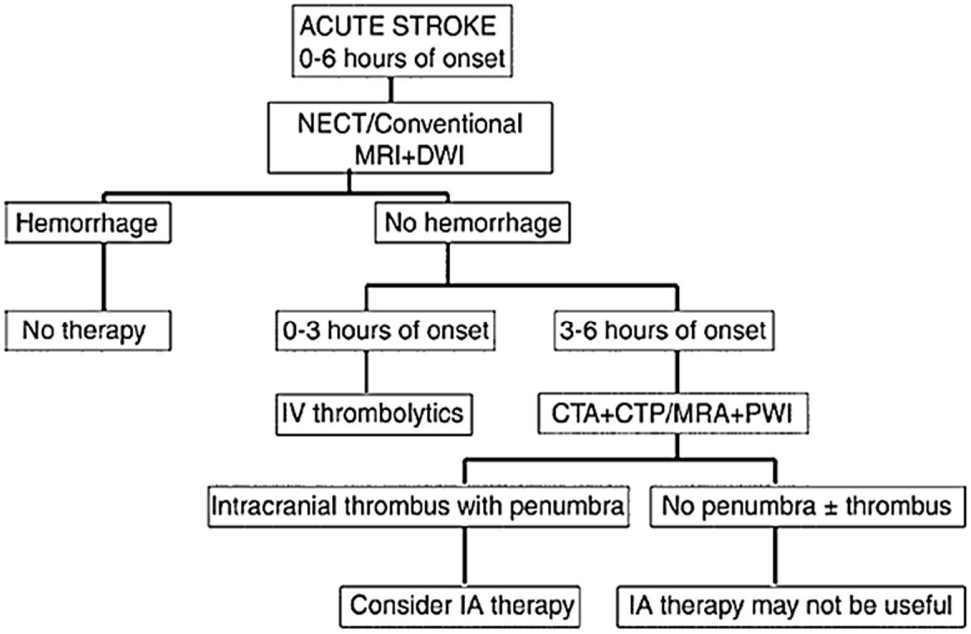
❖ 5 Basic Goals Of Acute Stroke Imaging

- Differentiate between hemorrhagic and ischemic stroke.
- Identification of the location and extent of intravascular clot
- The presence and extent of "ischemic core" (irreversibly damaged tissue)
- The presence and extent of "penumbra" (hypoperfused tissue at risk for infarction).
- Early identification of the aetiology or mechanism of the stroke (e.g. carotid atherosclerotic disease, vascular dissection, or others).

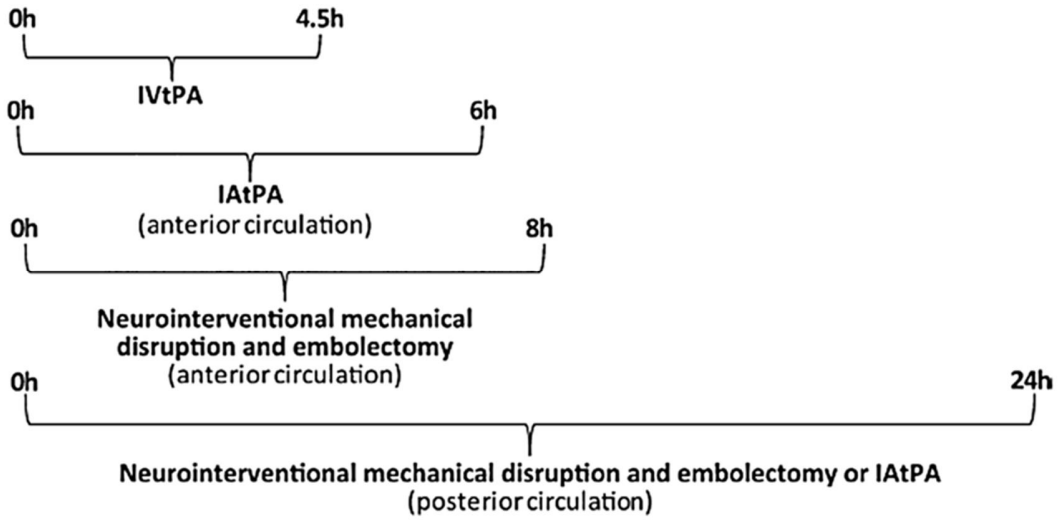
❖ 4Ps Goals Of Acute Stroke Imaging

Parenchyma	Assess early signs of acute stroke, rule out hemorrhage
Pipes	Assess extracranial circulation (carotid and vertebral arteries of the neck) and intracranial circulation for evidence of intravascular thrombus
Perfusion	Assess cerebral blood volume, cerebral blood flow, and mean transit time
Penumbra	Assess tissue at risk of dying if ischemia continues without recanalization of intravascular thrombus

❖Therapeutic Window In Early Treatment Of Hyperacute Cerebral Ischemia:



Time Windows*



* This is a rough guideline, and if there is a DWI/PWI mismatch, penumbra, or positive DWI/negative FLAIR combination, some believe intervention may still be warranted

❖ Causes of Ischemic Stroke:

1) Thromboembolic:
   - Cardiac embolism
     - Atrial fibrillation
     - ventricular aneurysm
     - endocarditis
   - Paradoxical embolism
   - Atherosclerotic embolism
   - Fat embolism
   - Air embolism
2) Atherosclerotic
3) Dissection
4) Hypoxic-Ischaemic-Injury (HII)

❖ Risk Factors of Ischemic Stroke:

1) Age: Advancing age
2) Sex: Males – Postmenopausal females
3) Family history
4) Smoking
5) Hyperglycemia
6) Hyperlipidemia
7) Hypertension
8) Homocystinuria

Imaging Protocol in stroke

❖ CT VS MRI in Acute Stroke

MR Imaging versus CT for Acute Stroke

MR Imaging	Computed Tomography
Less widely available than CT outside of major stroke centers	Widespread access
Contraindications (eg, electronic implants, patient intolerance, medical instability)	Greater acquisition speed
Gradient-echo imaging superior to CT for the detection of acute hemorrhage	Highly sensitive for the exclusion or confirmation of hemorrhage
Diffusion-weighted imaging more sensitive than nonenhanced CT for the early detection of acute ischemia	Findings at CBV–perfusion CT and CT angiography–source imaging correlate with lesion size and ischemic lesion volumes at diffusion-weighted imaging
Much more sensitive than nonenhanced CT for the detection of acute stroke	Multimodal CT survey (nonenhanced CT, perfusion CT, CT angiography) may be as sensitive as MR imaging
Comparison of findings at diffusion-weighted imaging and perfusion-weighted imaging represents the key fact of mismatch theory models	Mismatch can be determined with CBV (infarct core)–CBF (penumbra) maps
No radiation dose	Higher radiation dose

❖ 3 Imaging Strategies in Acute Stroke After NCCT:

1) Go to the angiography suite immediately after the initial NCCT.
2) CTA to assess vascular patency (after NCCT), with or without perfusion imaging, to better characterize the site of occlusion and the ischaemic tissue before making a decision about endovascular treatment.
3) MR imaging: T1, T2, FLAIR, DWI + MR perfusion (PWI) + MR angiography (MRA) + T2*-weighted gradient-echo imaging with susceptibility weighted imaging (Acquisition time = 20 min + radiologist´s interpretation =25 min)

❖ Role of NCCT

- Detection of hemorrhage
- Detection of ischemic signs of established infarction.
- Depiction of subtle early signs of infarction and ischemia
- Detection of other possible mimics of stroke (eg, neoplasm, arteriovenous malformation) that could be the cause of the neurologic deficit.

❖ Role of CTA

- Define the occlusion site.
- Depict arterial dissection.
- Grade collateral blood flow.
- Characterize atherosclerotic disease.

❖ Role of DWI

⇨ DWI has emerged as the most sensitive and specific imaging technique for acute infarct detection, far better than NCCT or any other MRI sequence.
⇨ Identify infarct size
⇨ Identify infarct site (deep or subcortical lesions)
⇨ Identify infarct age

NCCT	DWI
⇨ Low sensitivity (39%) ⇨ High specificity (100%)	⇨ High sensitivity (88 to 100%) ⇨ High specificity (95 to 100%)

❖ Role of T2*

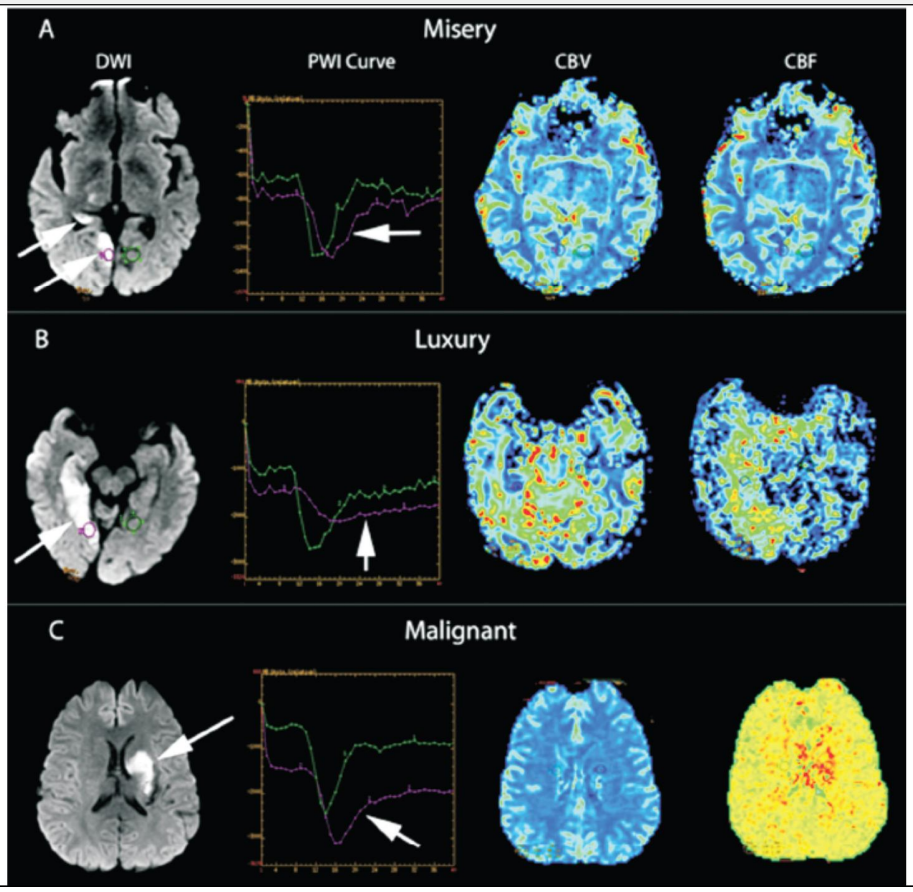
- Early detection of acute hemorrhage within 6 hours
- Identify previous microbleeds in acute ischemia
- Intravascular clot detection at the acute stage.
- Detection of Spontaneous Hemorrhagic Transformation Earlier Than CT
- Early Detection of Post-Thrombolytic Hemorrhagic Transformation.
- Diagnosis of Cerebral Venous Thrombosis

❖ MR Perfusion

Perfusion CT Analysis of Hyperacute Ischemic Stroke

	Analytic Tool		
Entity	MTT	CBF	CBV
Penumbra	Elevated (>145%)	Decreased	Normal or mildly increased
Infarct core	Elevated	Markedly decreased	Markedly decreased (<2.0 mL × 100 g^{-1})

Profile	Cerebral blood flow	Cerebral blood volume	Clinical interpretation
Misery profile	Decreased	Increased	Observed in ischaemic territories with poor collateral flow.
Luxury profile	Increased	Normal	Represents the relative hyperemia of critically ischaemic tissue following recanalization of regions with tissue acidosis.
Malignant profile	Decreased	Decreased	Represents a state of near-zero perfusion; it is the usual finding in the "core" region.



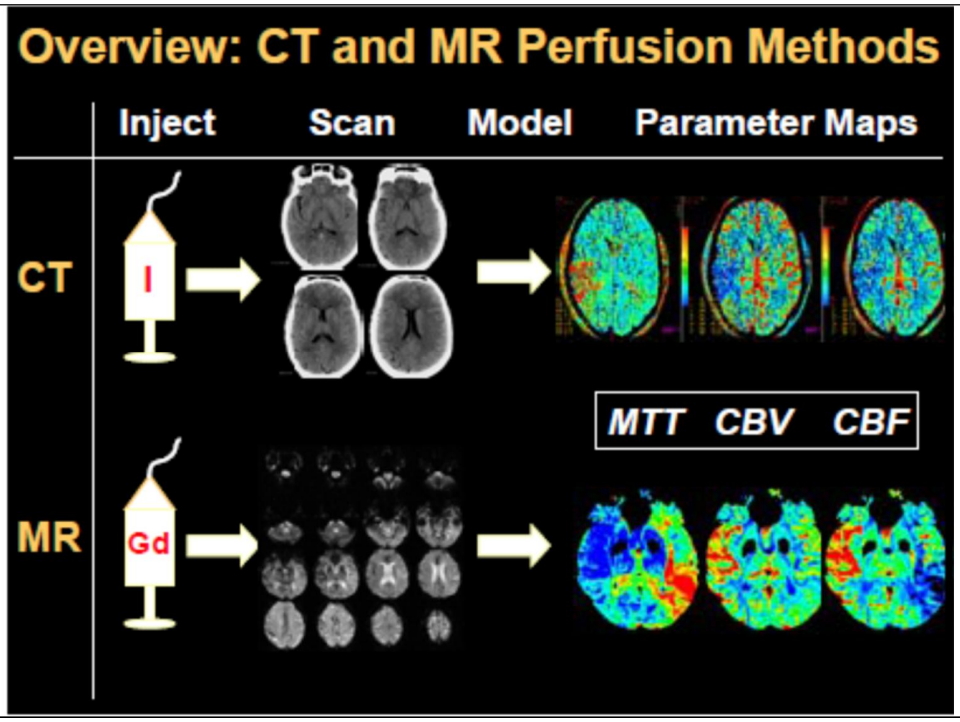
Key Perfusion Parameters

- **Cerebral Blood Flow (CBF)**
  - Delivery of blood to tissue / unit time
  - Units: ml / 100g brain / min
- **Cerebral Blood Volume (CBV)**
  - Measure of *autoregulation*
  - Units: ml / 100 g brain
- **Mean Transit Time (MTT)**
  - Average time to flow thru capillaries (artery → vein)
  - Units: seconds

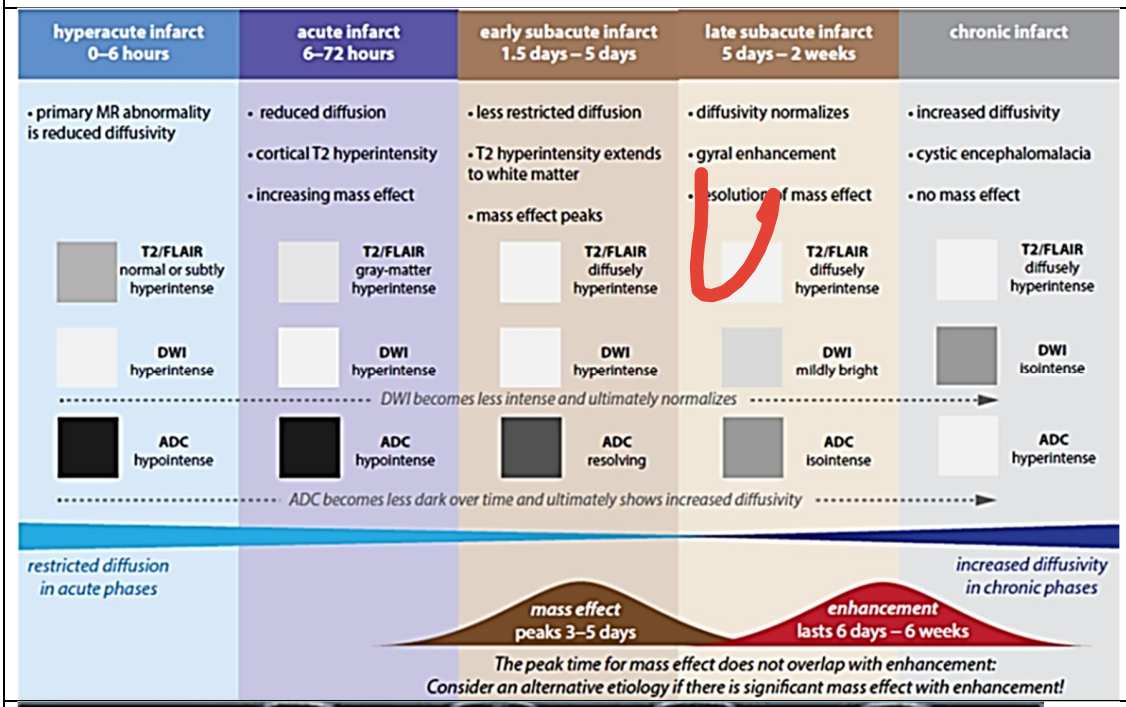
$$CBF = \frac{CBV}{MTT}$$

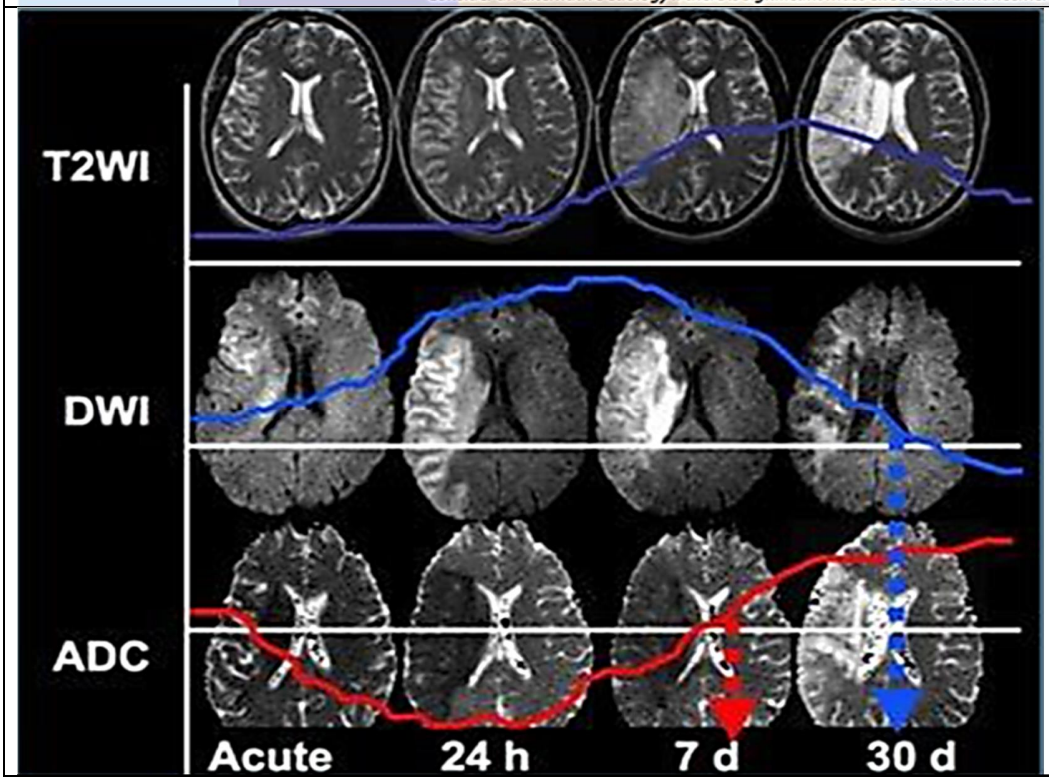
Central Volume Principle

Overview: CT and MR Perfusion Methods



❖ Evolution Of Infarction

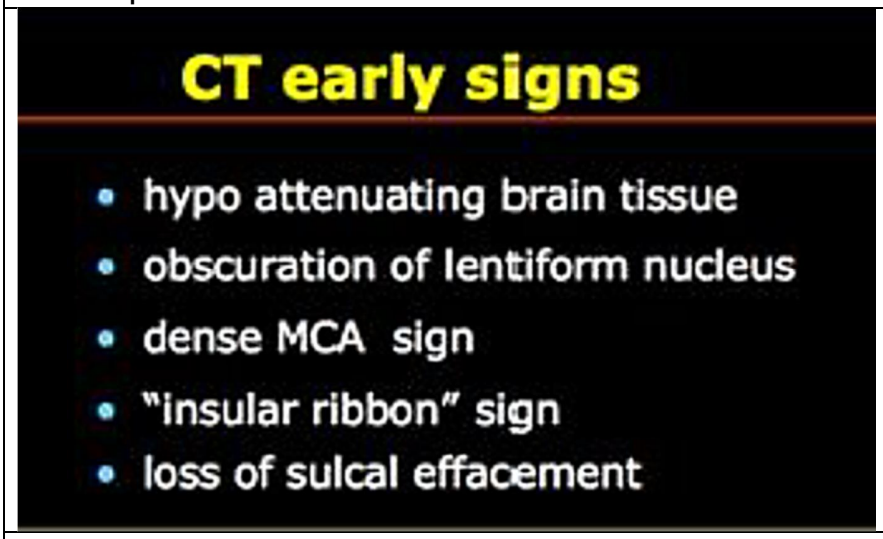

hyperacute infarct
0–6 hours
acute infarct
6–72 hours
early subacute infarct
1.5 days – 5 days
late subacute infarct
5 days – 2 weeks
chronic infarct
• primary MR abnormality is reduced diffusivity
• reduced diffusion
• cortical T2 hyperintensity
• increasing mass effect
• less restricted diffusion
• T2 hyperintensity extends to white matter
• mass effect peaks
• diffusivity normalizes
• gyral enhancement
• resolution of mass effect
• increased diffusivity
• cystic encephalomalacia
• no mass effect
T2/FLAIR normal or subtly hyperintense
T2/FLAIR gray-matter hyperintense
T2/FLAIR diffusely hyperintense
T2/FLAIR diffusely hyperintense
T2/FLAIR diffusely hyperintense
DWI hyperintense
DWI hyperintense
DWI hyperintense
DWI mildly bright
DWI isointense
DWI becomes less intense and ultimately normalizes
ADC hypointense
ADC hypointense
ADC resolving
ADC isointense
ADC hyperintense
ADC becomes less dark over time and ultimately shows increased diffusivity
restricted diffusion in acute phases
increased diffusivity in chronic phases
mass effect peaks 3–5 days
enhancement lasts 6 days – 6 weeks
The peak time for mass effect does not overlap with enhancement:
Consider an alternative etiology if there is significant mass effect with enhancement!



I- Hyperacute Infarct (1st 6hrs)

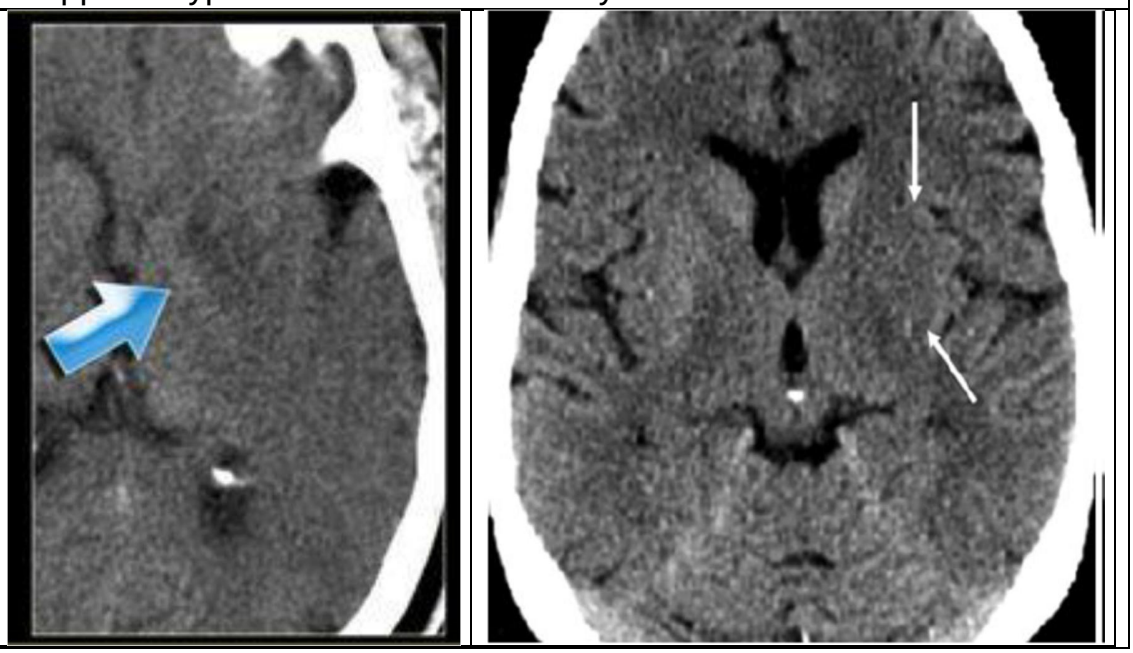
A) Non-contrast CT

- Noncontrast CT is the initial test of choice to exclude hemorrhage.
- Noncontrast CT is relatively insensitive to detect early infarction compared to MRI



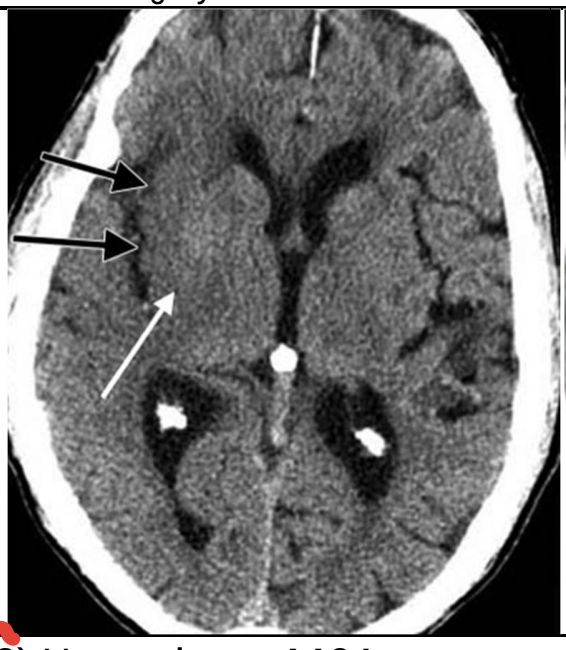
1) Obscuration of Lentiform Nucleus

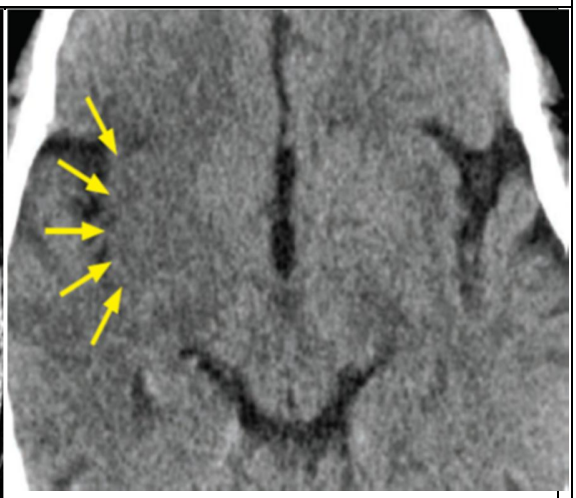
- Appears hypoattenuated because of cytotoxic edema.



2) Insular Ribbon Sign

- Loss of gray–white differentiation in the insula.





3) Hyperdense MCA

- Represent acute intravascular thrombus

3D CT Angiography

B) MRI:

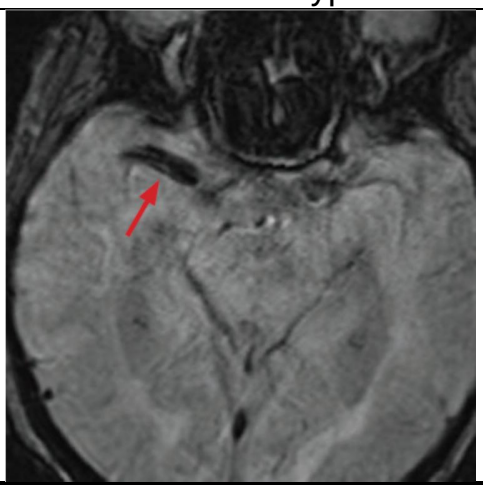
1) Gradient Image

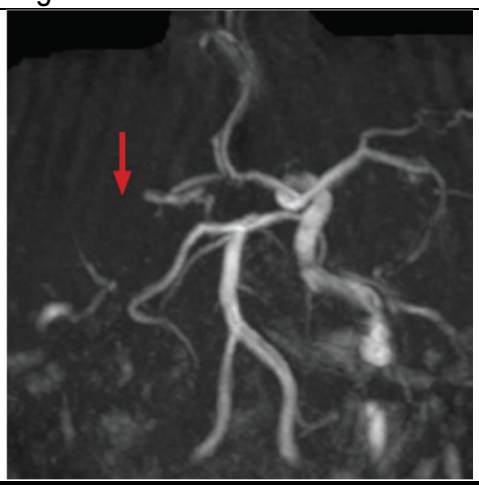
- Blooming artifact in the right MCA (red arrow).

2) MRA

- Complete occlusion of the right MCA (red arrow)

- Represents intraluminal thrombus
- correlate to the hyperdense artery sign that can be seen on CT.



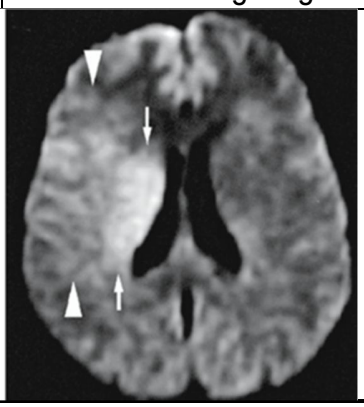


3) T2/FLAIR

Normal

4) DWI

Restricted = High signal

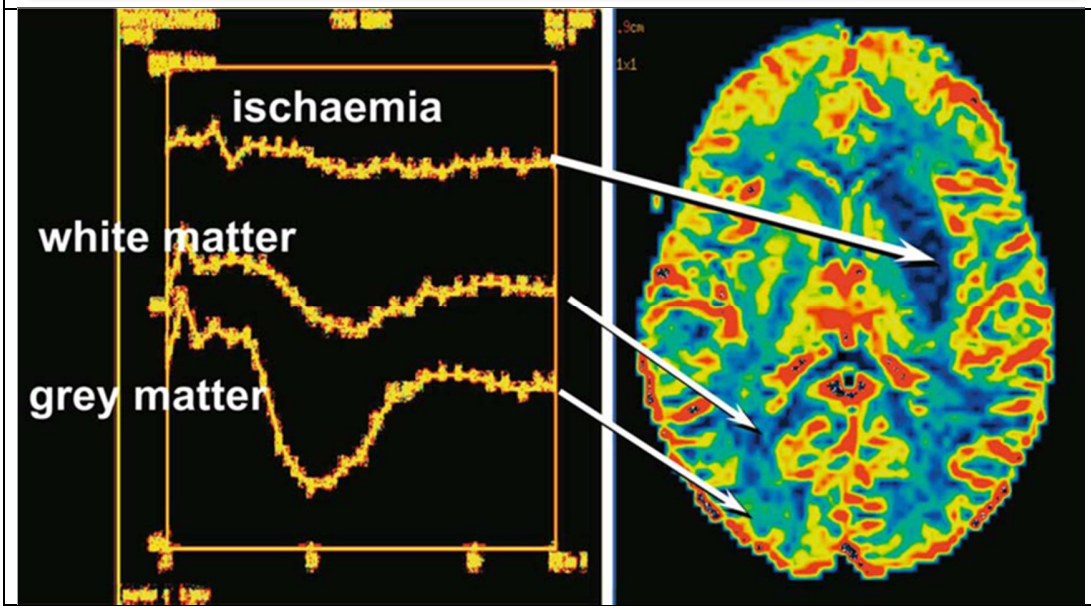


5) ADC

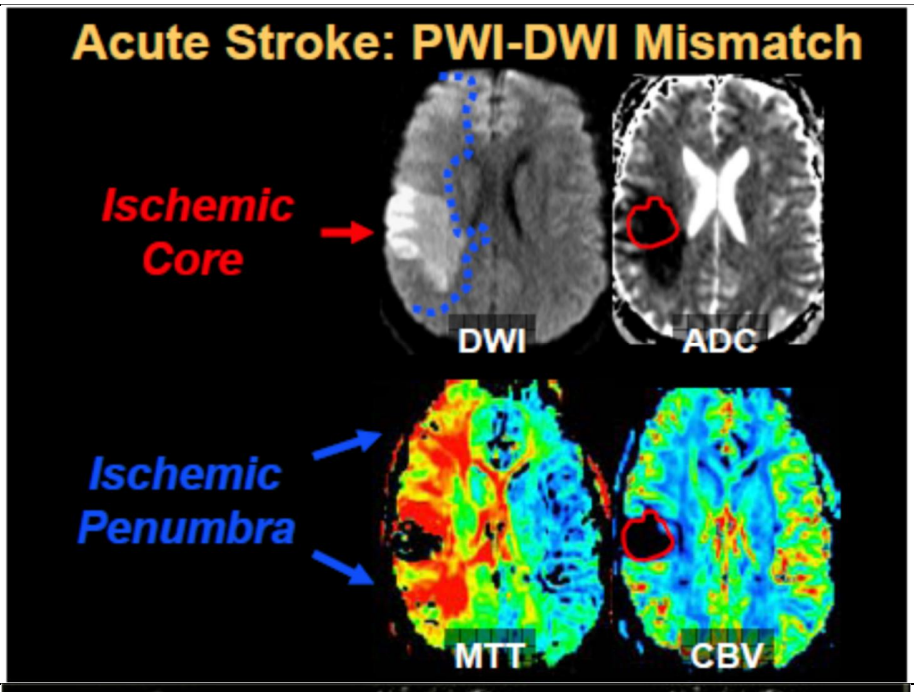
Low signal

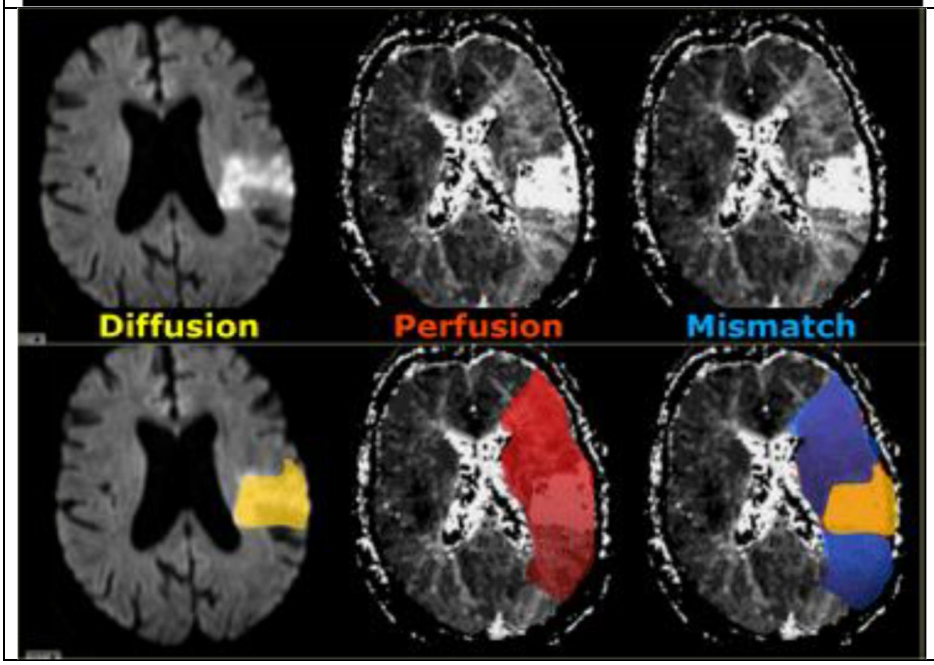
❖ CT / MR Perfusion

Entity	MTT	CBF	CBV
Penumbra	Elevated (>145%)	Decreased	Normal or mildly increased
Infarct core	Elevated	Markedly decreased	Markedly decreased (<2.0 mL x 100 g^{-1})



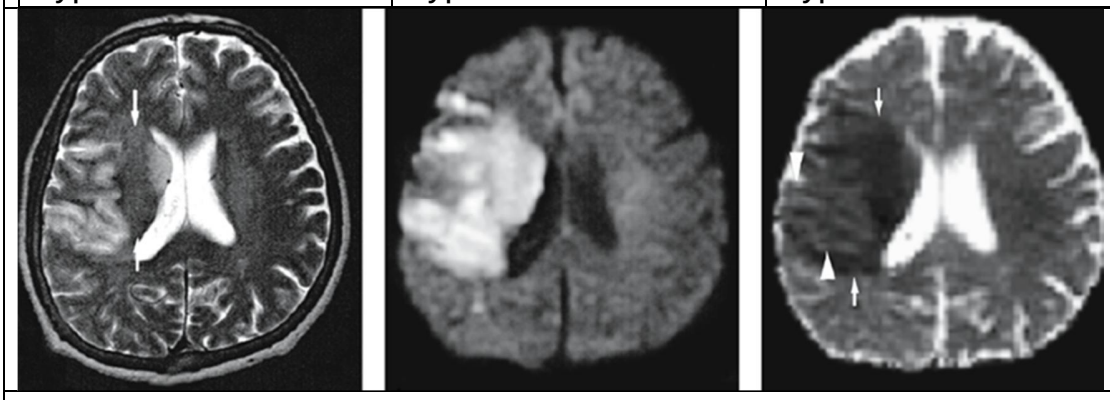
❖ Diffusion – Perfusion Mismatch





II- Acute Infarct (6hrs – 3 days)

T2/FLAIR	DWI	ADC
Hyperintense	Hyperintense	Hypointesne



❖ Mass effect

- due to surrounding vasogenic oedema due to blood-brain barrier breakdown
- Effacement of the overlying cortical sulci
- Compression of the adjacent ventricle
- The mass effect peaks at 3–4 days

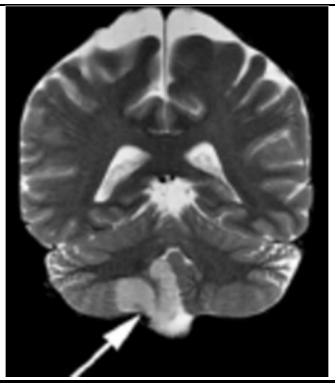
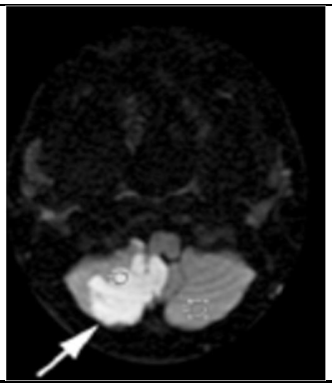
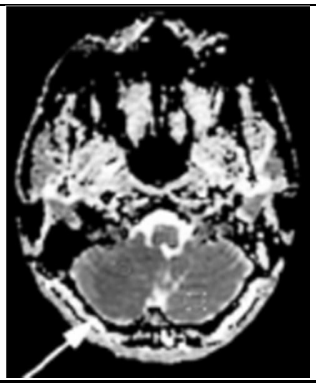
❖ Contrast Enhancement:

- Some arterial enhancement, due to increased collateral flow.

❖ Perfusion images

- Increase in size of the infarct core
- Decrease in size of the penumbra.

III- Early Subacute Infarct (1.5 days–5 days)

❖T2 / FLAIR	❖DWI	❖ADC
Marked hyperinternse GM & WM	Restricted = High signal	Less dark or iso
		

❖ Perfusion

- Continued expansion of the infarct core
- further reduction in the ischemic penumbra.

❖ Mass effect

- due to surrounding vasogenic oedema due to blood-brain barrier breakdown
- Effacement of the overlying cortical sulci
- Compression of the adjacent ventricle
- The mass effect peaks at 3–4 days

❖ Contrast Enhancement:

- Meningeal enhancement
- Blood flow to the affected brain is re-established by leptomeningeal collaterals and ingrowth of new vessels into the region of infarction

IV- Late Subacute Infarct (5 days–2 weeks)

❖DWI & ADC

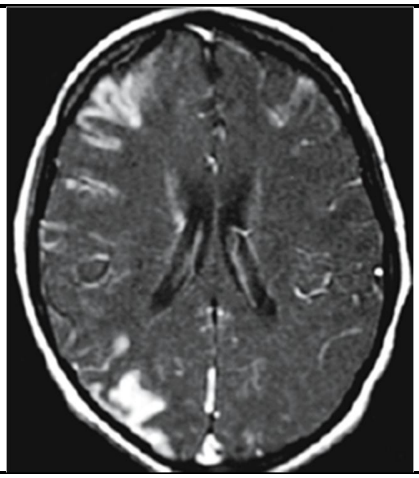
- DWI neutralize or may remain bright due to underlying T2 shine through
- ADC map will either return to normal or hyperintense

❖Mass Effect:

- Resolution of vasogenic edema and reduction in mass effect.

❖Gyral Enhancement (Luxury Perfusion)

- A subacute infarction will not typically demonstrate both mass effect and enhancement simultaneously (unlike tumor)
- "2-2-2" rule: Enhancement begins at 2 days, peaks at 2 weeks, and disappears by 2 months.



❖ Haemorrhagic Transformation

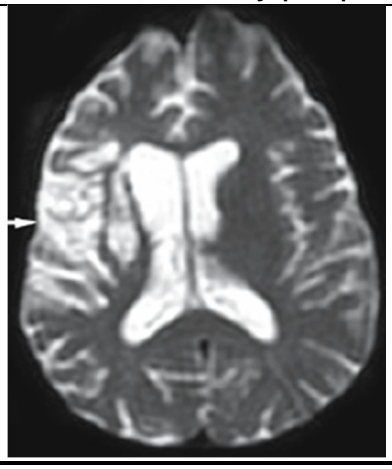
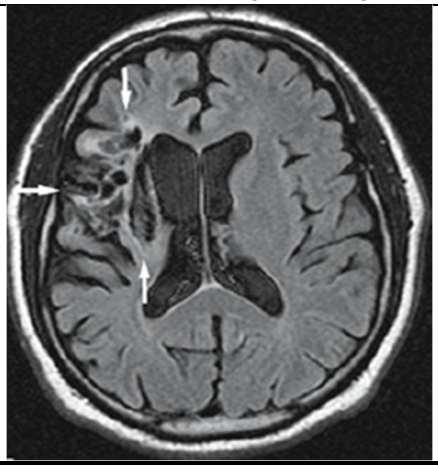
V- Chronic Infarct

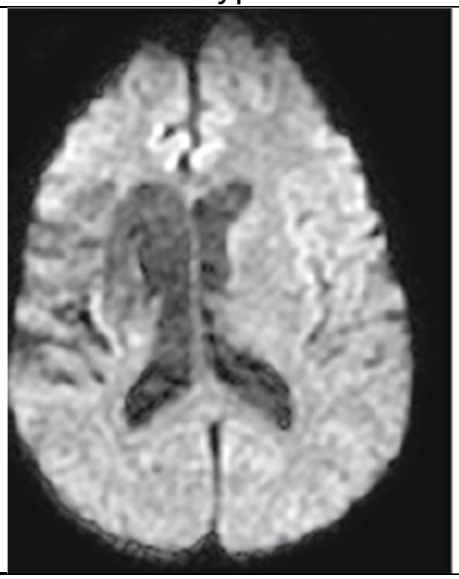
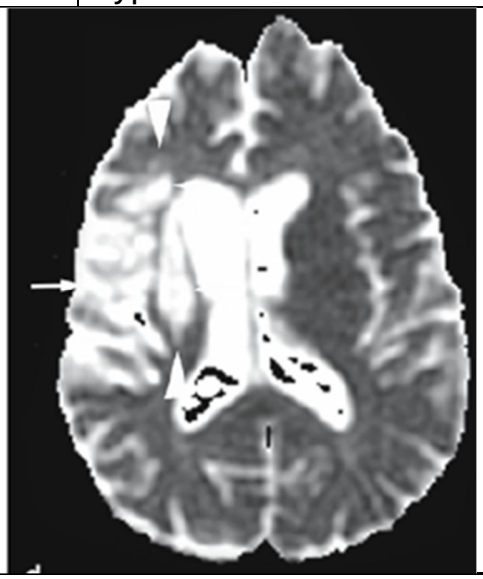
❖Cystic Encephalomalacia & Gliosis

- Infarction, cellular debris and dead brain tissue are removed by macrophages and replaced by cystic encephalomalacia and gliosis.

❖Negative mass effect:

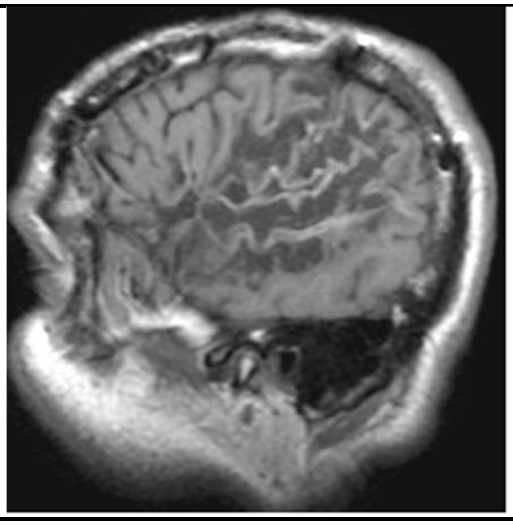
- Widening of the overlying cortical sulci
- Exvacudilatation of the adjacent ventricle.

❖ T2	❖ FLAIR
• Cystic encephalomalacia: High T2 and low FLAIR signal • Surrounded by peripheral mild high T2 and FLAIR signal of gliosis	
	

❖ DWI	❖ ADC
Facilitated = Hypointense	Hyperintense
	

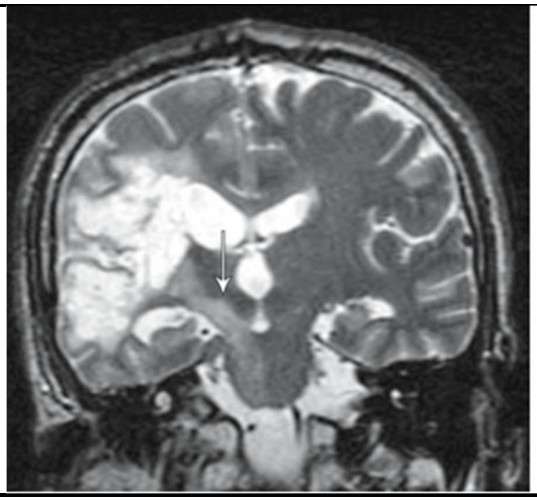
❖ Cortical Laminar Necrosis

- Characterized by deposition of lipidladen macrophages after ischemia
- T1 and T2 hyperintensity



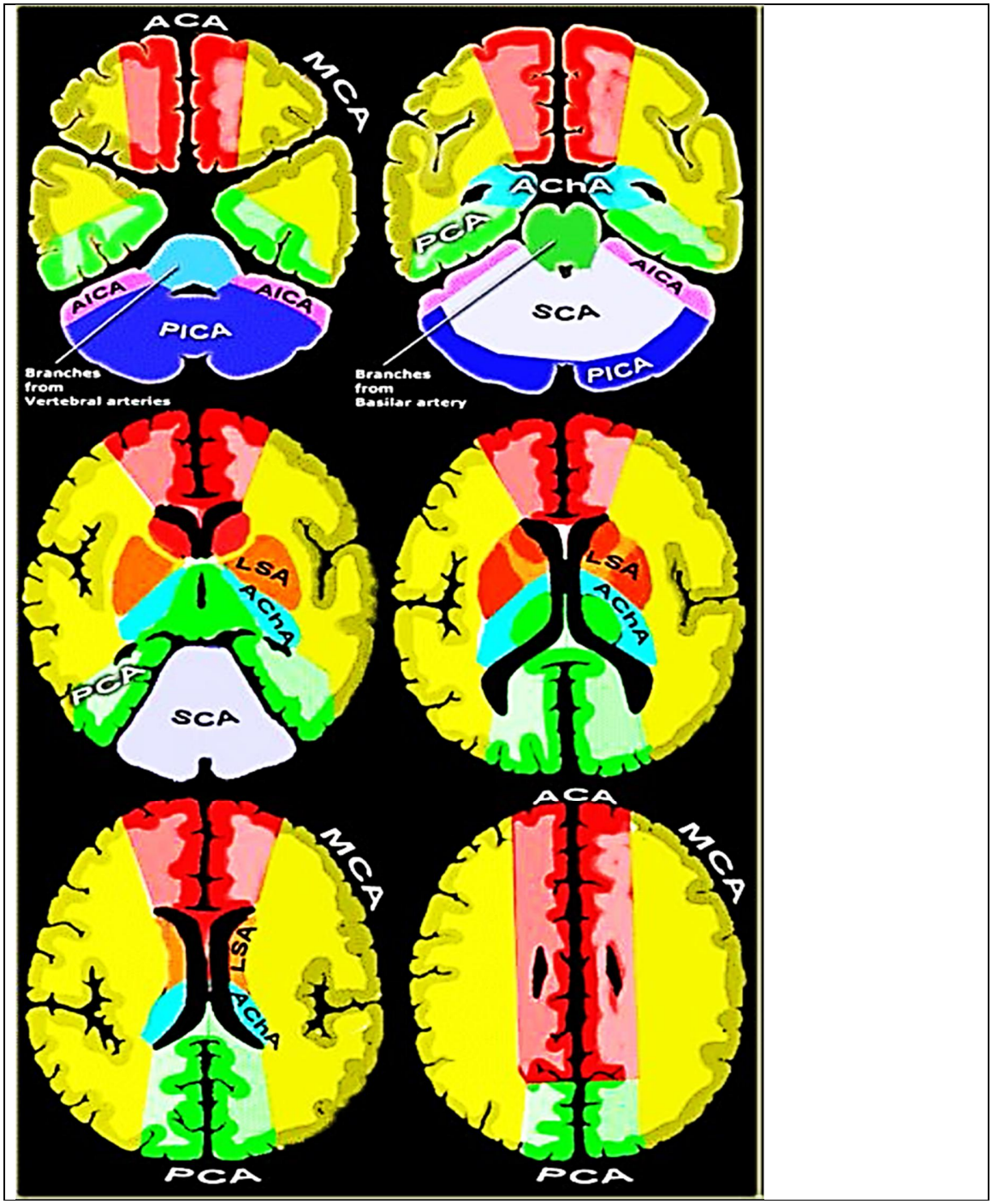
❖Wallerian Degeneration

- T2/FLAIR: Atrophy + Hyperintensity of ipsilateral corticospinal tract along:
    - o Posterior column of internal capsule
    - o Cerebral peduncle of mid brain
    - o Ventral pons



Crossed Cortical Dysgenesis

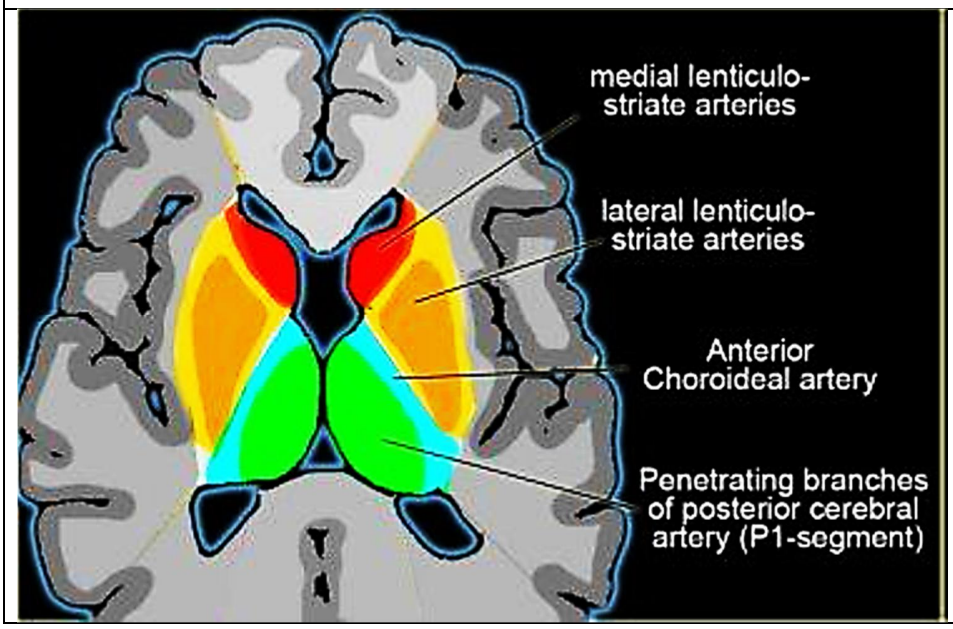
Arterial Territories



Anterior Cerebral Artery (ACA in red) • Cortical branches of ACA supply the medial part of the frontal and the parietal lobe and anterior portion of the corpus callosum • The deep penetrating medial LSA-branches (discussed later).
Middle cerebral artery (MCA in yellow) • Cortical branches of the MCA supply the lateral surface of the hemisphere, except for the medial part of the frontal and the parietal lobe (anterior cerebral artery), and the inferior part of the temporal lobe (posterior cerebral artery). • The deep penetrating lateral LSA-branches (discussed later).
Posterior cerebral artery (PCA in green) • Cortical branches of the PCA supply the inferomedial part of the temporal lobe, occipital pole, visual cortex, and splenium of the corpus callosum. • Posterior thalamoperforating arteries (discussed later)
Superior Cerebellar Artery (SCA in grey) • Supply the superior and tentorial surface of the cerebellum.
Anterior Inferior Cerebellar Artery (AICA in purple) • Supply the lateral surface of the cerebellum .
Posterior Inferior Cerebellar Artery (PICA in blue) • Supply the inferior occipital surface of the cerebellum.
Branches from vertebral and basilar artery • Supply the medulla oblongata (in blue) and the pons (in green).

❖ Critical Small Branches Arising From The Circle Of Willis

- ICA (C6 segment)
  - ⇨ Anterior choroidal artery => supply the optic chiasm, hippocampus, and posterior limb of the internal capsule.
- A1 segment
  - ⇨ Recurrent artery of Heubner => supply the caudate head + Anterior limb of internal capsule.
  - ⇨ Medial lenticulostriate perforators => supply medial basal ganglia: Caudate + Anterior limb of internal capsule.
- M1 segment
  - ⇨ Lateral lenticulostriate perforators => supply the lateral basal ganglia: Lateral putamen, external capsule + posterior limb of internal capsule
- Pcom
  - ⇨ Anterior thalamoperforators => supply the thalamus.
- P1 segment
  - ⇨ Posterior thalamoperforators => supply the thalamus.
  - ⇨ Artery of Percheron is a variant where there is a dominant thalamic perforator supplying the ventromedial thalami bilaterally and the rostral midbrain.


medial lenticulo-striate arteries
lateral lenticulo-striate arteries
Anterior Choroideal artery
Penetrating branches of posterior cerebral artery (P1-segment)

Vascular Territory Infarction

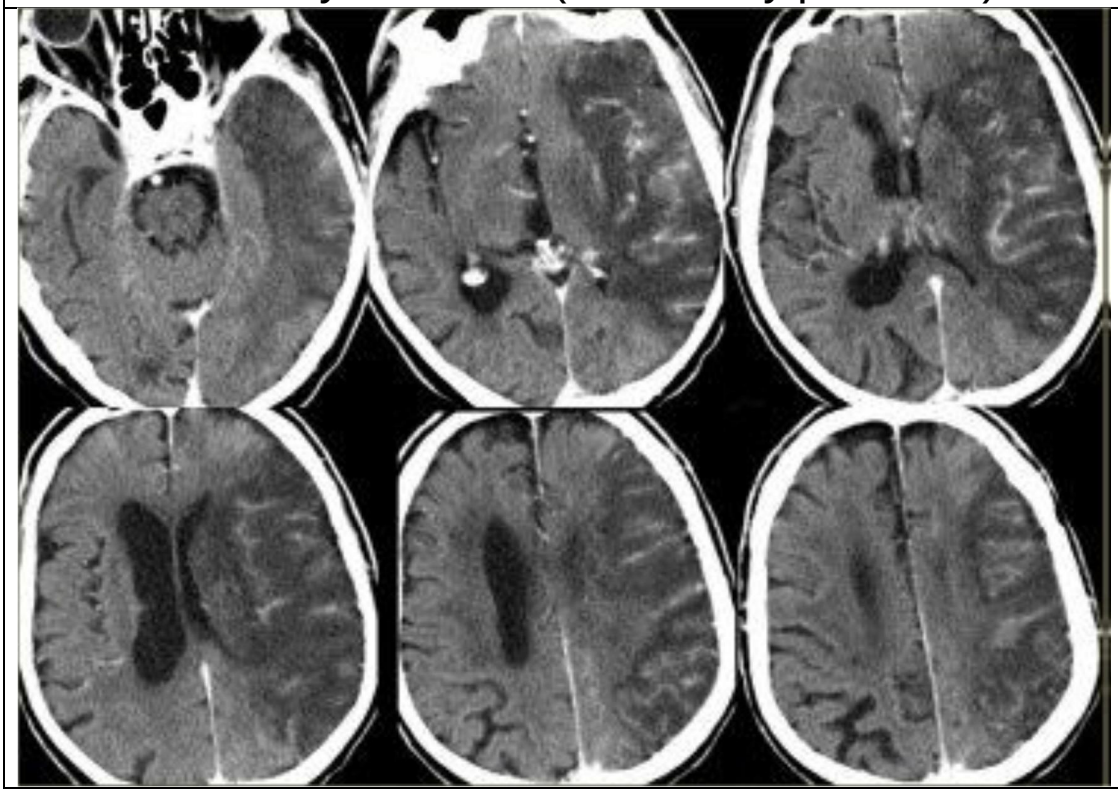
1) MCA Territory Infarction

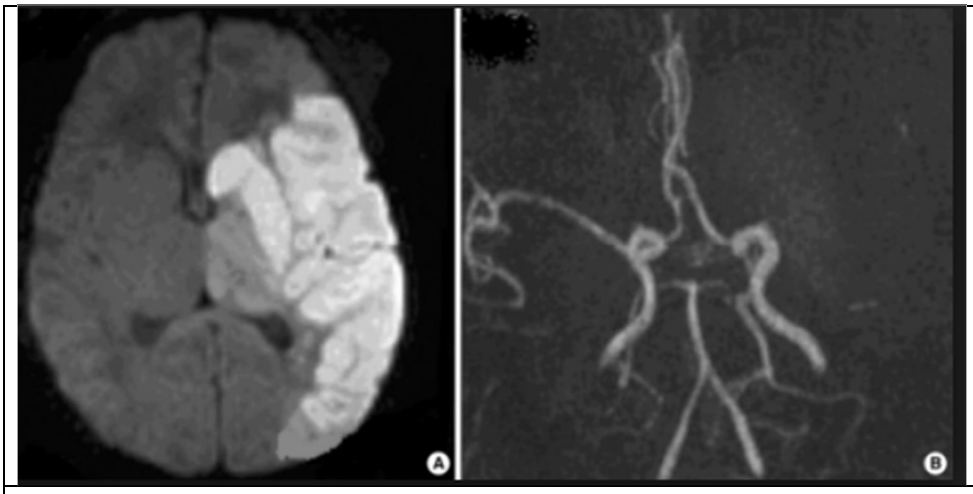
- Most commonly affected territory in a cerebral infarction, due to the size of the territory and the direct flow from internal carotid artery into the middle cerebral artery, providing the easiest path for thromboembolism.

Clinical Picture:

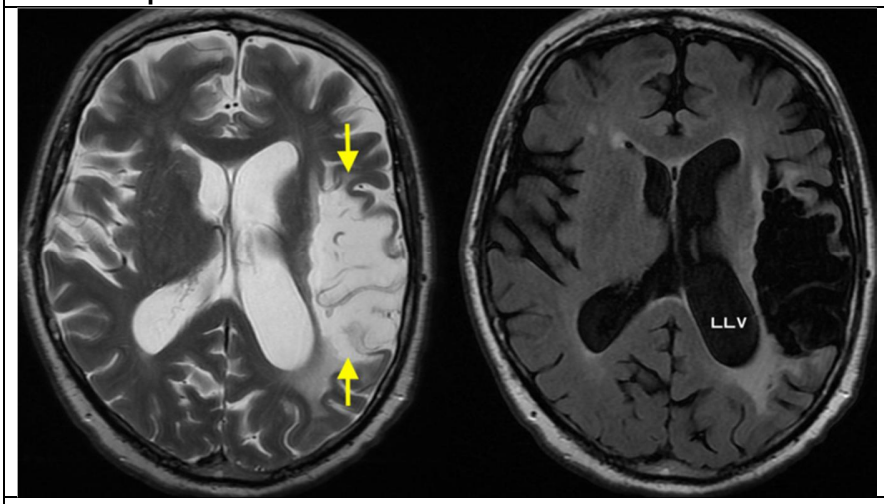
- Contralateral hemiparesis
- Contralateral hemisensory loss
- Hemianopia
- Aphasia: if the dominant hemisphere is involved.

❖ MCA Territory Infarction (with luxury perfusion)



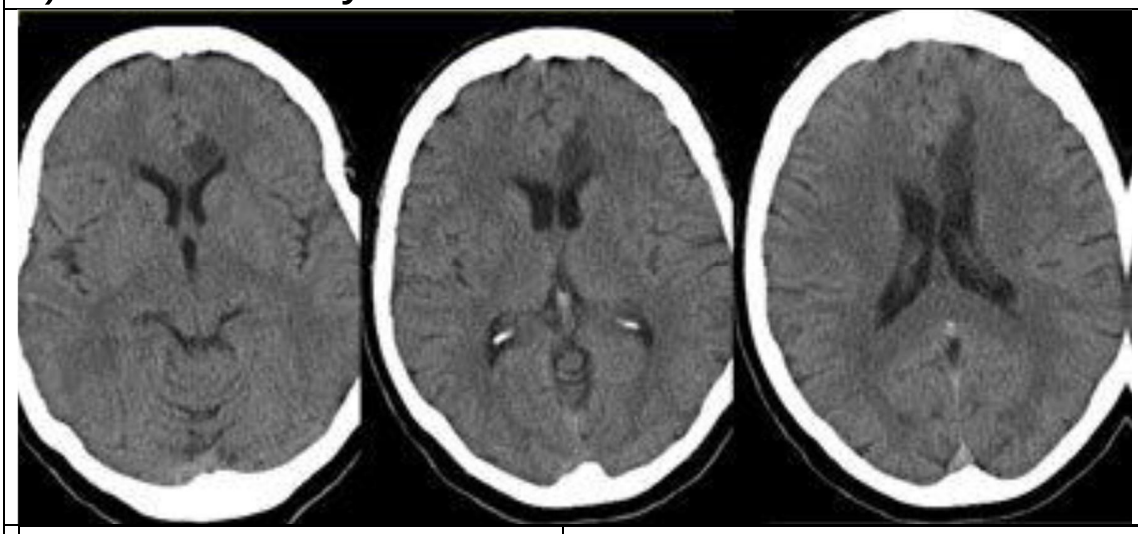


❖ Encephalomalacia due to old MCA infarct

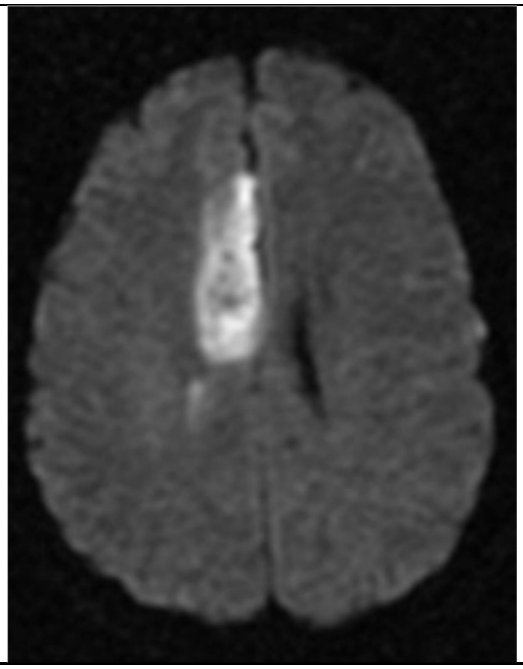


❖ LSA Hemorrhagic Infarction

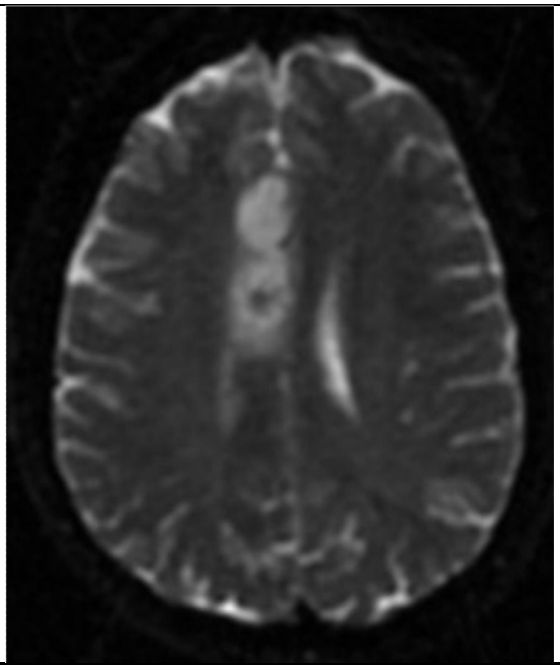
2) ACA Territory Infarction



Axial DWI

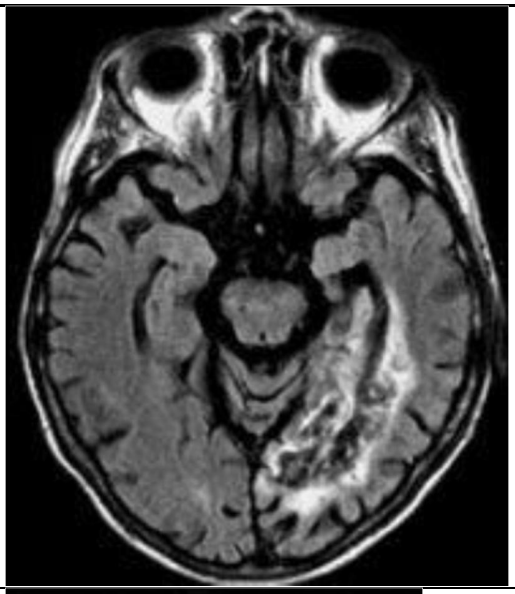
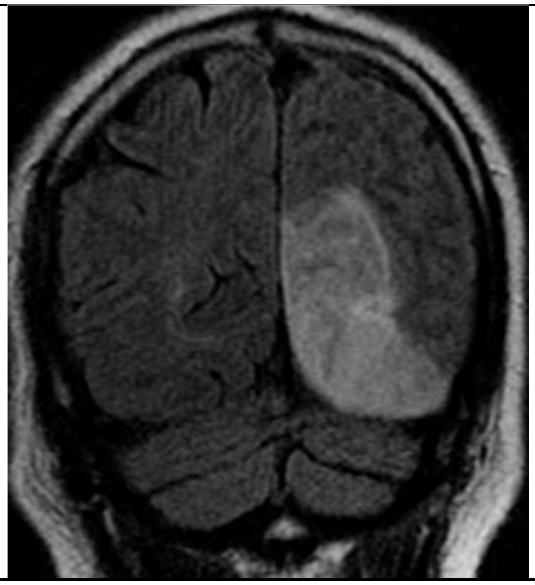


Axial ADC



3) PCA Territory Infarction

- Visual and sensory deficits.
- less overall chronic disability than those with anterior cerebral, middle cerebral, or basilar artery infarctions

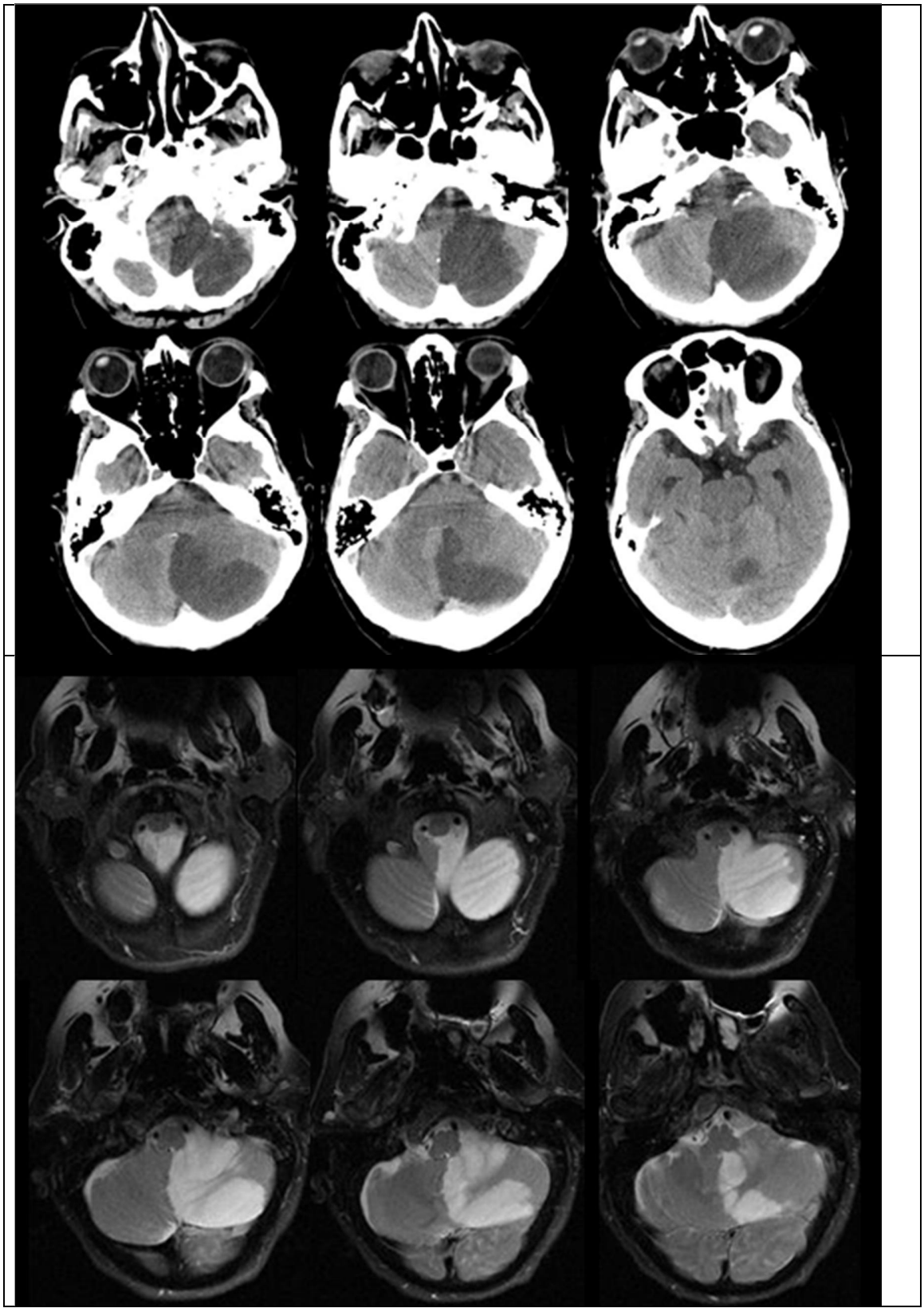



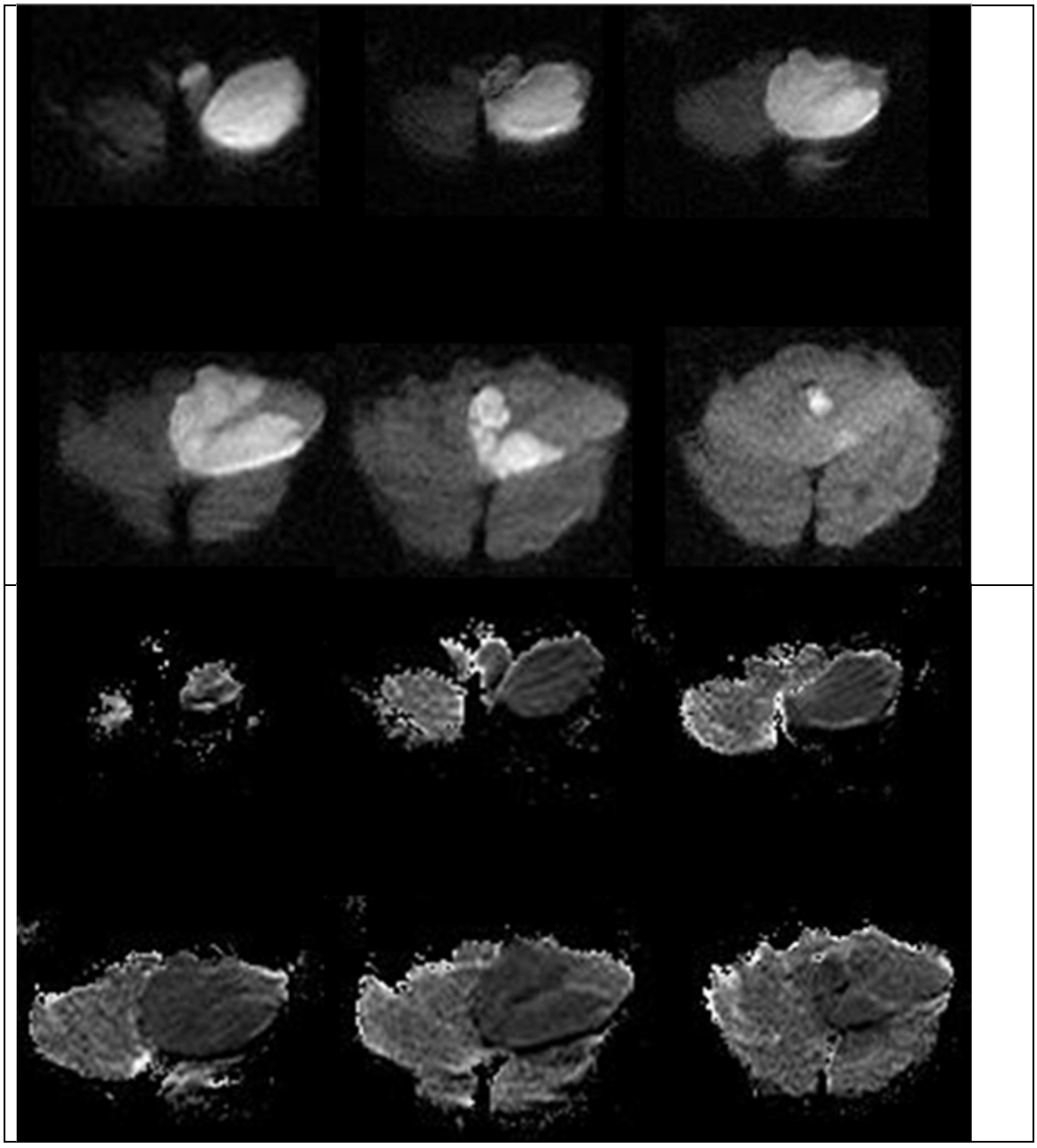
4) SCA Infarction

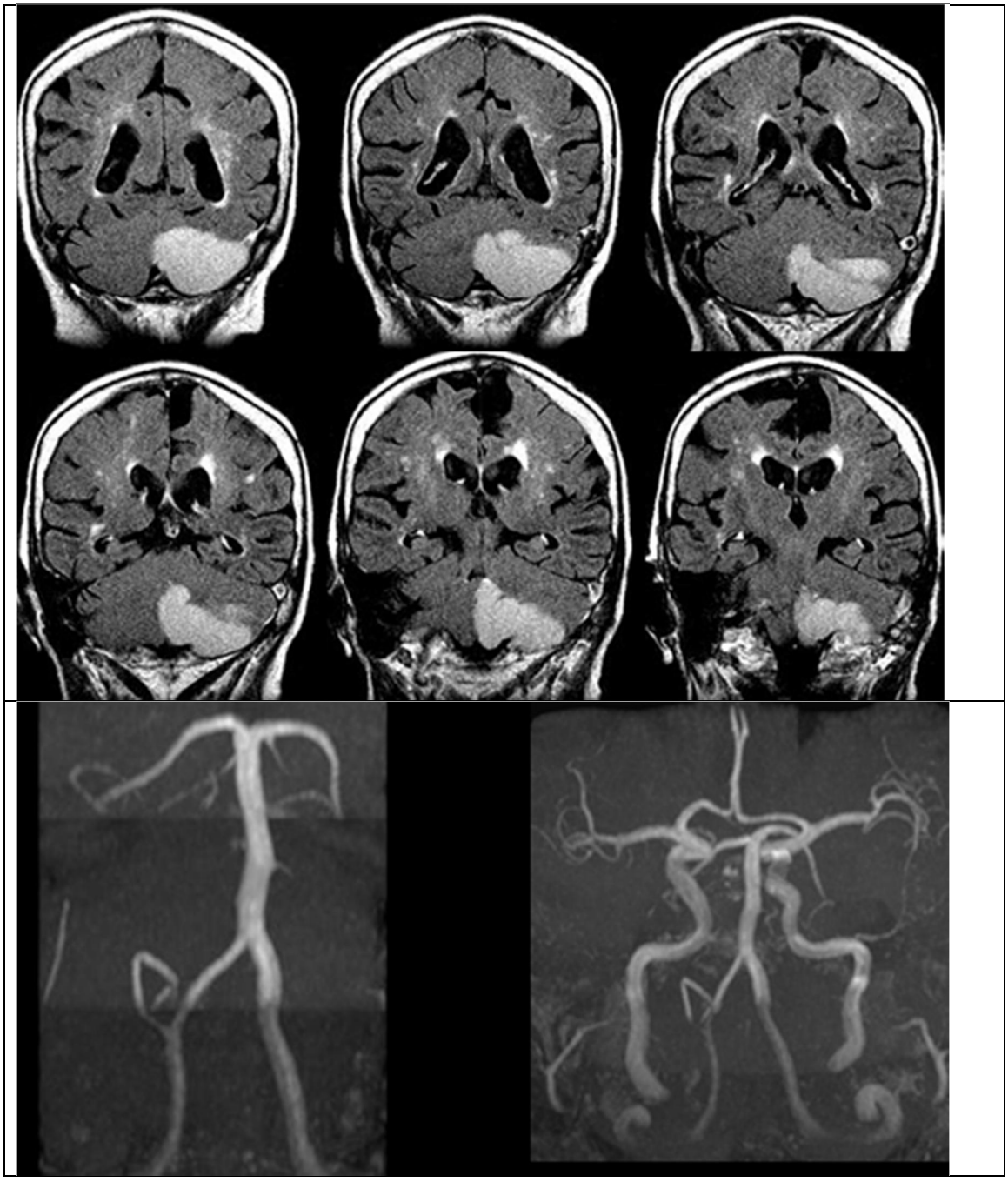
5) PICA Territory Infarction

Lateral Medullary (Wallenberg) Syndrome:

- is the most commonly obstructed cerebellar artery.
- Ipsilateral face => loss of pain and temperature sensation
- Contralateral body => loss of pain and temperature sensation
- Ipsilateral Horner's syndrome
- Ataxia, dysphagia, vertigo, nystagmus, hiccups







6) Moya – Moya Disease

Associated with:

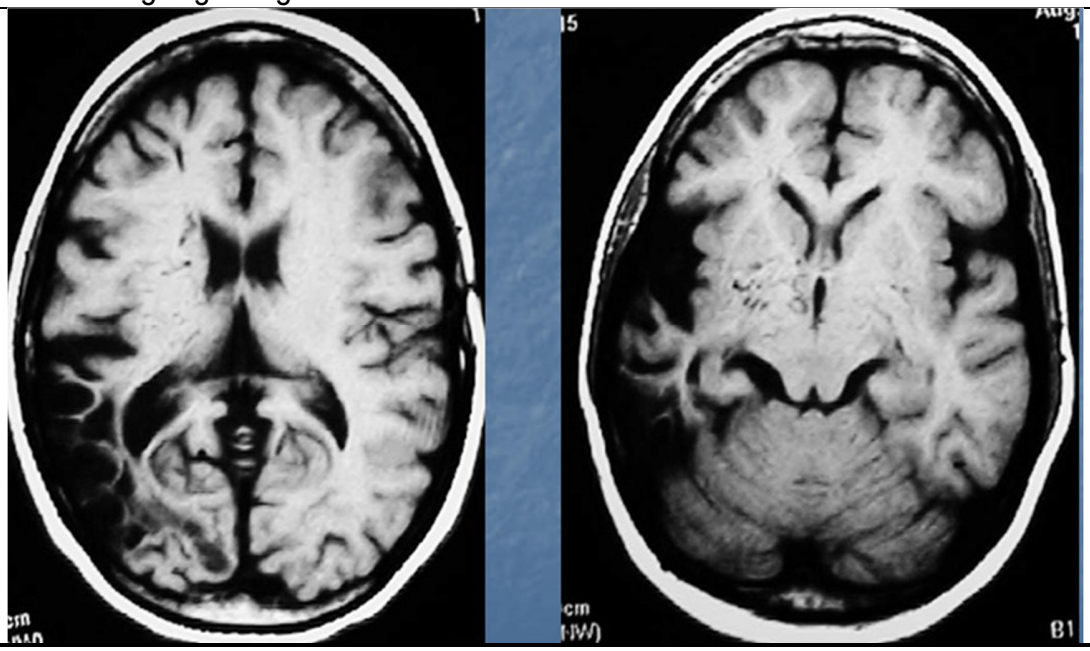
- ❖ Head and neck infections
- ❖ Tuberous sclerosis – Fanconi's anemia
- ❖ Irradiation

Angiography:

- Occlusion of supraclinoid ICA
- Lack of normal MCA
- Tangle of fine collaterals (puff of smoke):
  - o Lenticulostriate
  - o Thalamoperforators
  - o Choroidal arteries

MRI

- MCA territory infarct / atrophy
- Basal ganglia signal void collaterals



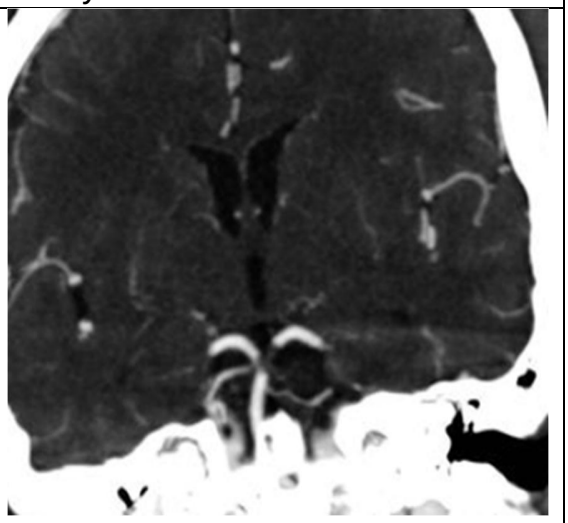
7) Top of Basilar Artery Syndrome = Rostral Brainstem Infarction

- Thromboembolic occlusion of the top of the basilar artery.

⇨ Bilateral thalamic ischaemia due to occlusion of perforator vessels.

Clinical picture:

- Visual and oculomotor deficits
- Behavioral abnormalities
- Somnolence, hallucinations and dreamlike behaviour
- Motor dysfunction is often absent

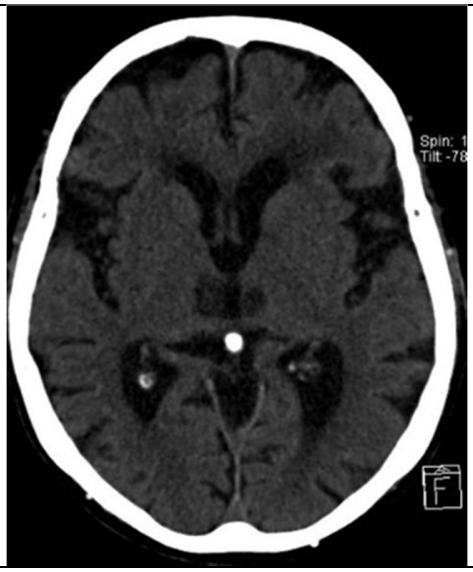
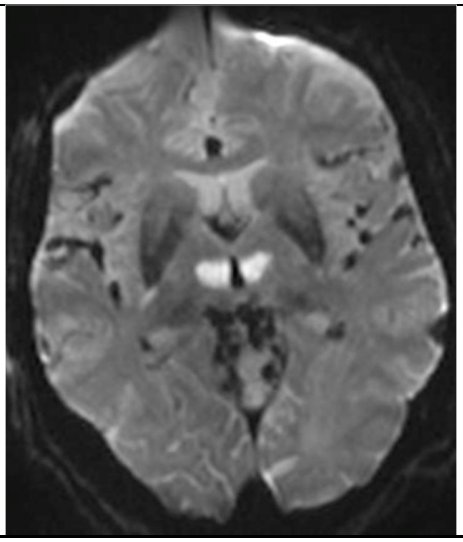
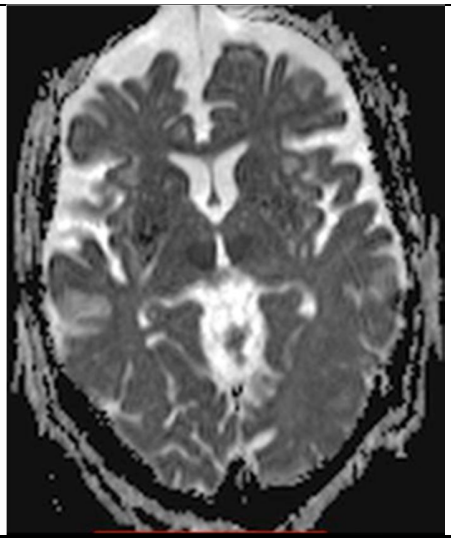
NCCT: hyperdense tip of basilar artery	CTA: occlusion of tip of basilar artery
	

8) Artery of Percheron Territory Infarct = Paramedian Thalamic Syndrome

- Rare variant of the posterior cerebral circulation characterised by a solitary arterial trunk that supplies blood to the paramedian thalami and the rostral midbrain bilaterally

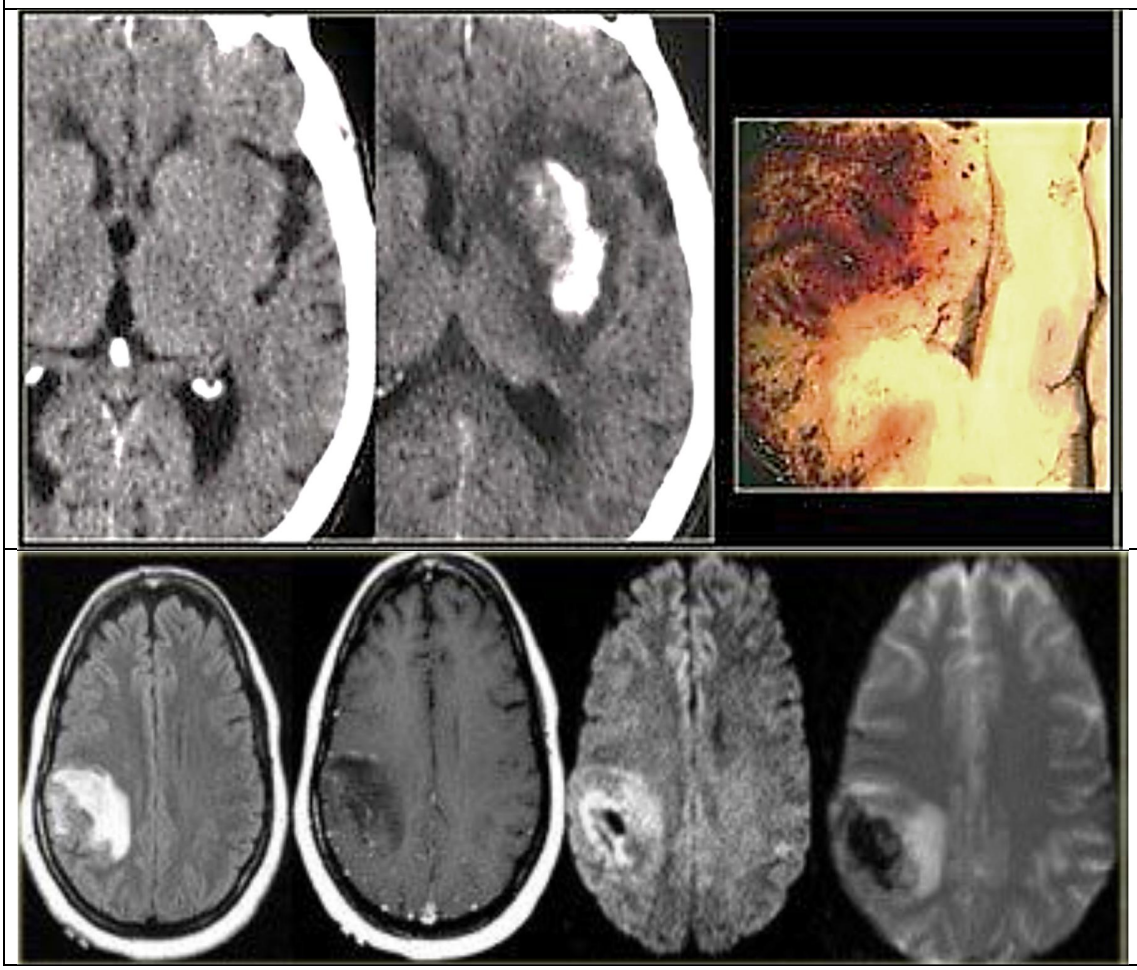
- Bilateral infarcts areas involving the rostral midbrain and/or ventromedial thalamus.

- Triad of altered mental status, vertical gaze palsy, and memory impairment

Axial FLAIR	NCCT
	
Axial DWI	**Axial ADC**
	

Hemorrhagic Infarcts

- 15% of MCA infarcts are initially hemorrhagic.
- Hemorrhage is most easily detected with non-contrast CT
- But it can also be visualized with gradient echo



Watershed Infarcts

- Watershed infarcts occur at the border zones between major cerebral arterial territories as a result of hypoperfusion.

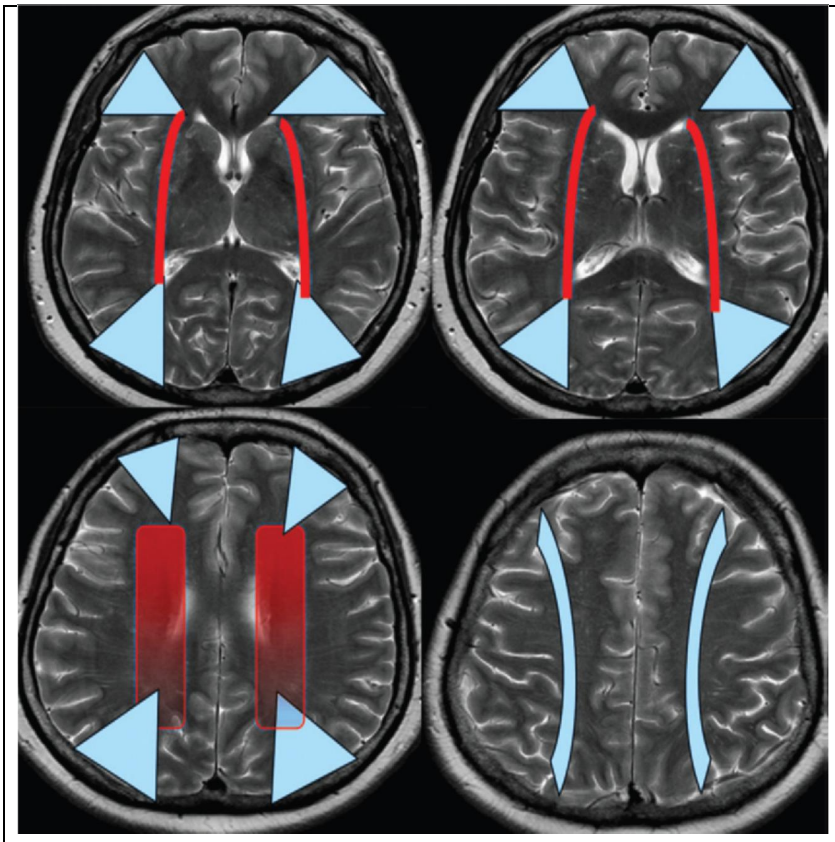
❖ 2 Patterns Of Border Zone Infarcts:

1) External = Cortical Border Zone Infarctions

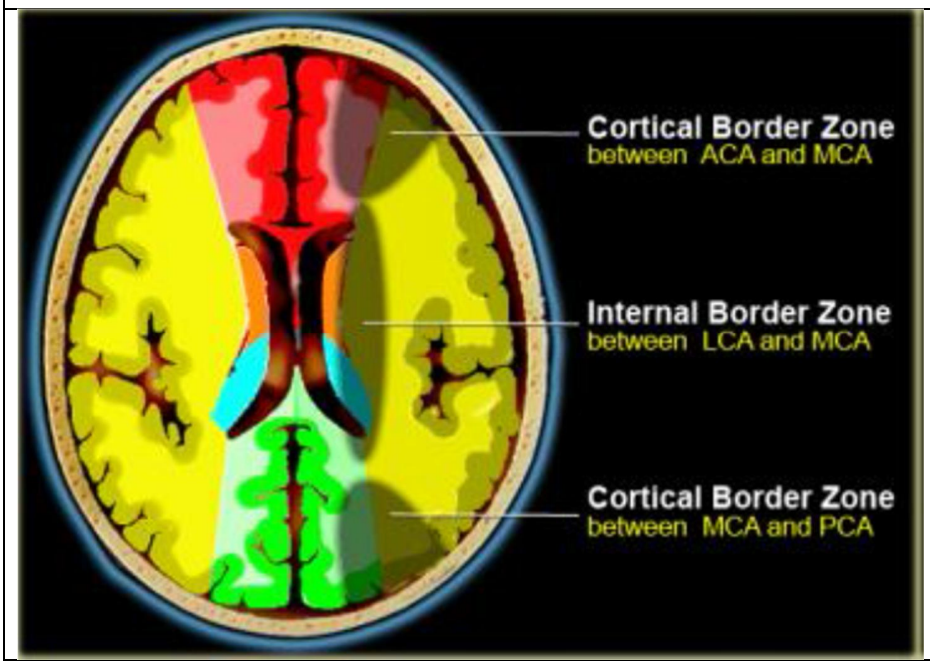
- Embolic in nature; microemboli from the heart or atherosclerotic plaques in major arteries
- Less frequently associated with hemodynamic compromise.
- Wedge shaped cortical infarcts.
- Frontal = between ACA & MCA
- Posterior parieto-occipital = between MCA & PCA
- Parasagittal fronto-parietal = between ACA & MCA

2) Internal = Subcortical Border Zone Infarctions

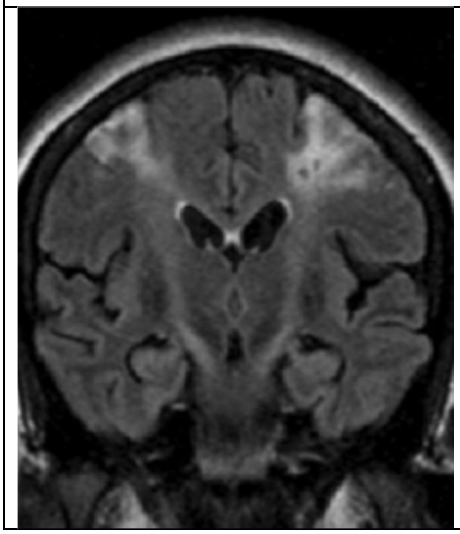
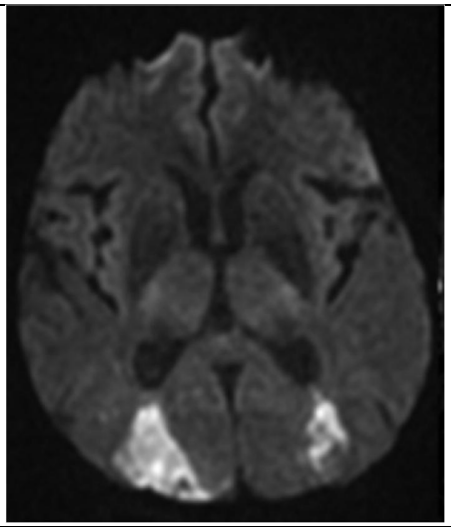
- Caused mainly by arterial stenosis or occlusion, or hemodynamic compromise.
- Represent the "tip of the iceberg" of decreased perfusion reserve and may be predictive of impending stroke.
- Multiple small rosary beads or cigar shaped infarcts
- Infarctions of the deep white matter of the centrum semiovale and corona radiata parallel to the adjacent ventricle
- The border zone between lenticulostriate perforators and the deep penetrating cortical branches of the MCA
- The border zone of deep white matter branches of MCA and ACA.



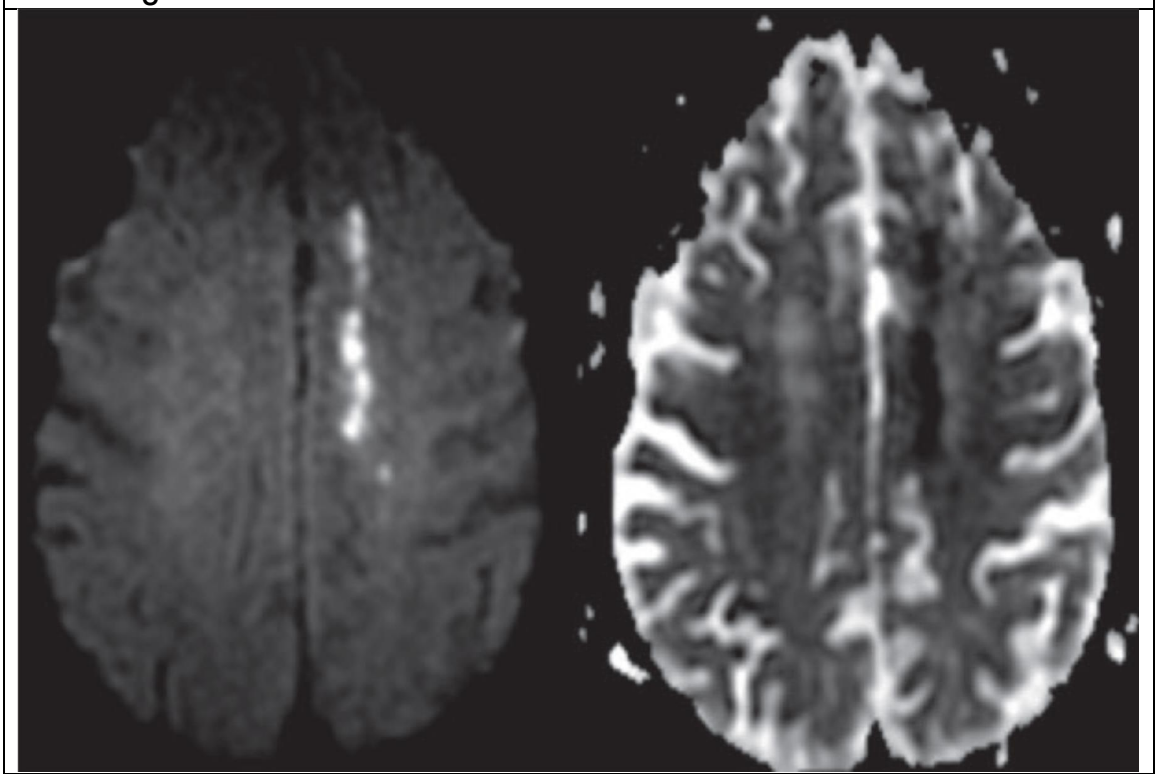
Blue = External Red = Ineternal



Cortical Border Zone Infarcts

Frontal = between ACA & MCA	Posterior parieto-occipital = between MCA & PCA
	

- Multiple internal border zone infarcts in a rosary like pattern along the left centrum semiovale.



Lacunar Infarcts

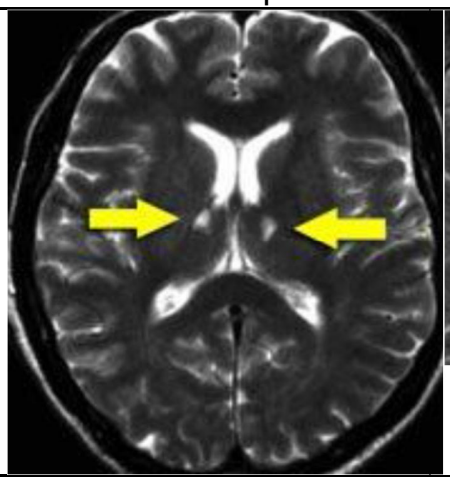
- Lacunar infarcts are small infarcts in the deeper parts of the brain (basal ganglia, thalamus, white matter) and in the brain stem.

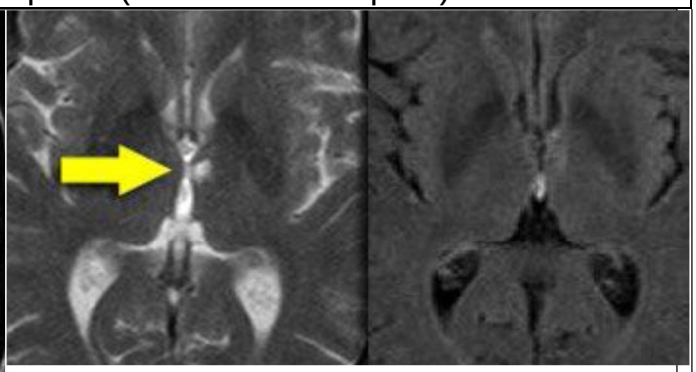
- Lacunar infarcts account for 25% of all ischemic strokes.
- Atherosclerosis is the most common cause of lacunar infarcts followed by emboli.

- Lacunar infarcts are caused by occlusion of deep penetrating artery:
  - Lenticulostriate arteries
  - Thalamoperforators
  - Brain stem perforators

Imaging findings:

- Small rounded / ovoid shaped lesion
- < 1cm
- T2 / FLAIR: high signal
- FLAIR: sometimes lacunar infarct is hardly seen
- DD: Dilated perivascular spaces (Virchow-Robin space)





Hypoxic-Ischemic Encephalopathy

HIE is a major cause of Death and Cerebral Palsy of children.

❖ Causes of HIE

A) Intrauterine Asphyxia:

Placental blood flow and gas exchange is interrupted:

- Fetal factors (fetomaternal hemorrhage, fetal thrombosis, and fetal bradycardia),
- Inadequate placental perfusion (maternal hypotension, preeclampsia, chronic vascular disease, abruptio placenta),
- Impaired maternal oxygenation (asthma, pulmonary embolism, pneumonia, carbon monoxide poisoning, severe anemia), or
- Disrupted umbilical circulation (tight nuchal cord, cord prolapse).

B) Intrapartum factors

- Forceps delivery,
- Breech extraction,
- Umbilical cord prolapse,
- Abruptio placentae,
- Tight nuchal cord,
- Maternal fever

C) Postnatal Asphyxia

- Severe hyaline membrane disease, pneumonia, meconium aspiration,
- Congenital heart anomalies that cause neonatal pulmonary failure or hypotension

❖ 4 Patterns of HIE:

I. Mild to moderate hypoxia in preterm infants.
II. Severe hypoxia in preterm infants
III. Mild to moderate hypoxia in term infants
IV. Severe hypoxia in term infants.

I- Mild to Moderate Asphyxia in Preterm Infants

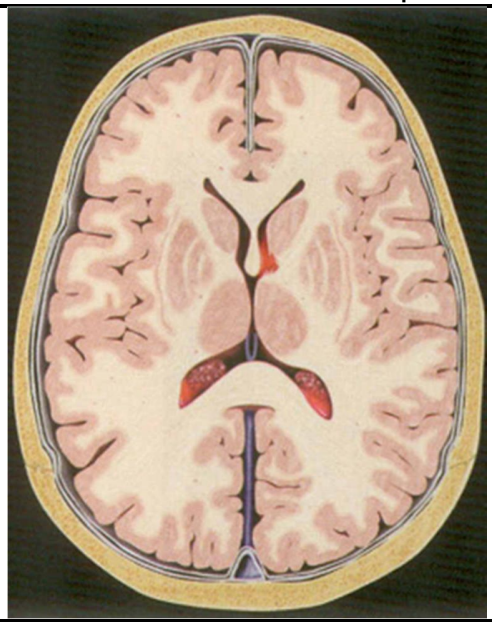
1) Germinal Matrix Hemorrhage (GMH):

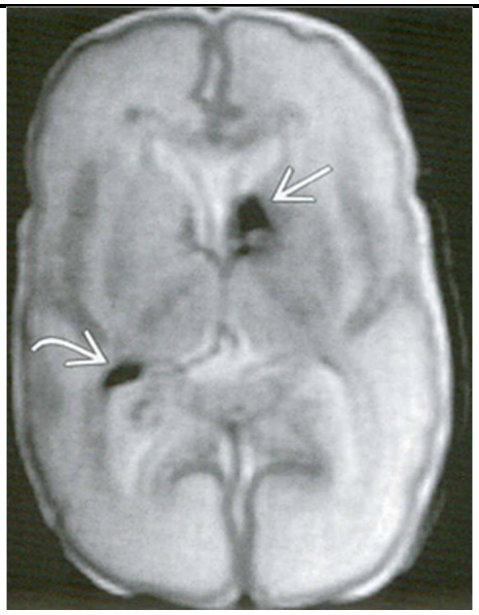
- usually occur during the first 24 hours of life.

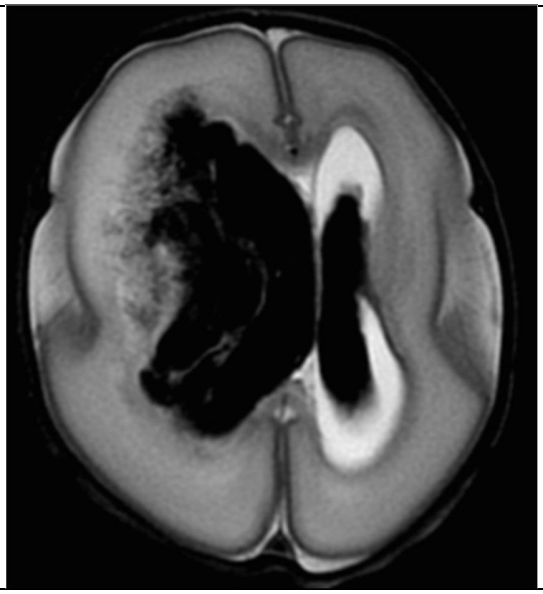
- Located anywhere in ventricular wall,
- most often caudothalamic notch = between the caudate nucleus and thalamus.
- may extend into the lateral ventricle(s) or periventricular brain parenchyma

• Grading of GMH:

- Grade 1: Subependymal hemorrhage = Confined to the caudothalamic groove
- Grade 2: Intraventricular hemorrhage without ventricular dilatation
- Grade 3: Intraventricular hemorrhage with ventricular dilatation
- Grade 4: Periventricular parenchymal venous infarction.

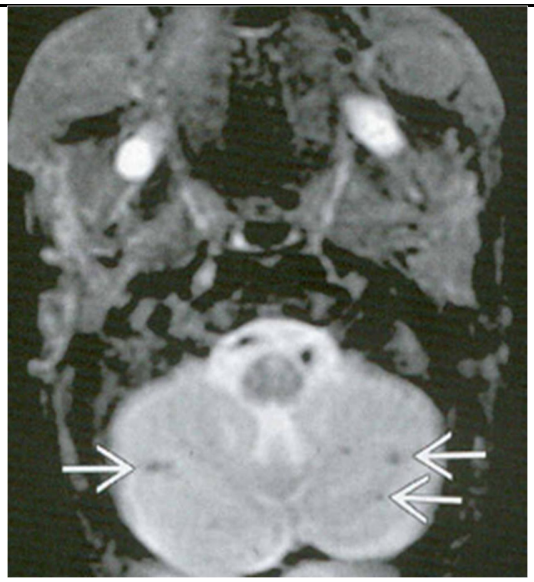






2) Cerebellar Hemorrhage:

- occur in approximately 25% of preterm infants with very low birth weight.
- They are assumed to arise from the germinal matrix zones in the external granule cell layer and the subependymal layer of the roof of the fourth ventricle.



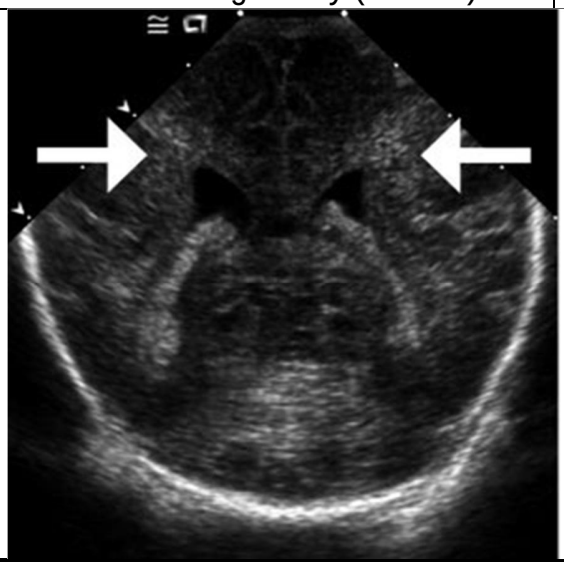
3) Periventricular Leukomalacia:

- WM injury may be focal (adjacent to frontal horns and trigones) or diffuse
- Areas that correspond to the watershed zones in periventricular white matter in the premature brain.

❖ US features of PVL

Table 2. Classification of PVL Based on Ultrasound

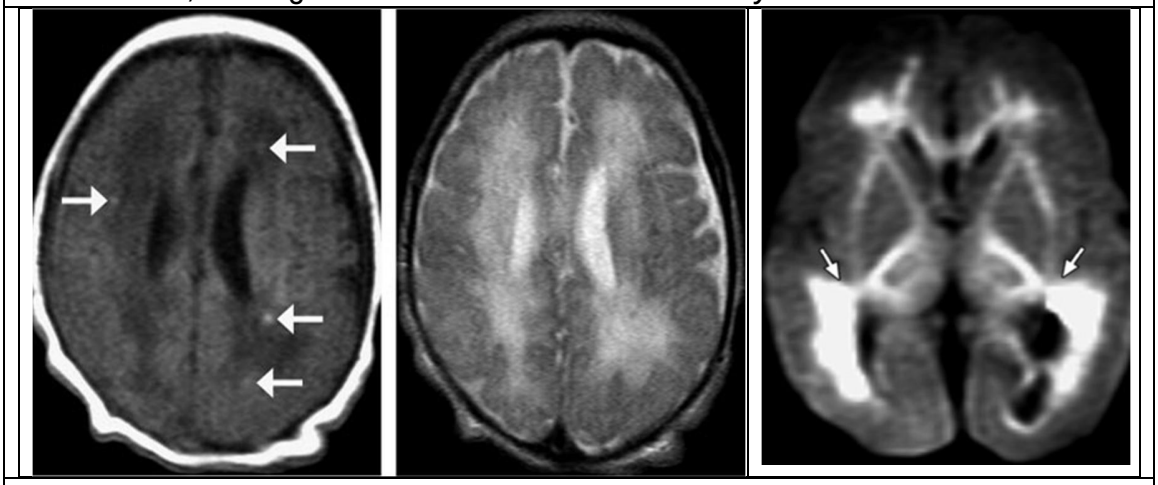
Grade I	Areas of increased echogenicity, usually seen within 24–48 hr after an insult and persisting beyond day seven, but not evolving into cysts
Grade II	Periventricular increased echogenicity evolving into small cysts, often located in the fronto-parietal periventricular white matter
Grade III	Periventricular increased echogenicity evolving into extensive cysts, particularly prominent in the parieto-occipital periventricular white matter. The cysts do not usually communicate with the lateral ventricles. They collapse after several weeks and are no longer visible on cranial ultrasound in two to three months. After several months, atrophy of the periventricular white matter leads to ventricular dilatation with irregular walls
Grade IV	Periventricular increased echogenicity evolve into extensive cysts which extend into the deep white matter

Periventricular white matter increased echogenicity (arrows).	Cystic changes + dilatation of the adjacent lateral ventricles
	

❖ MRI features of PVL

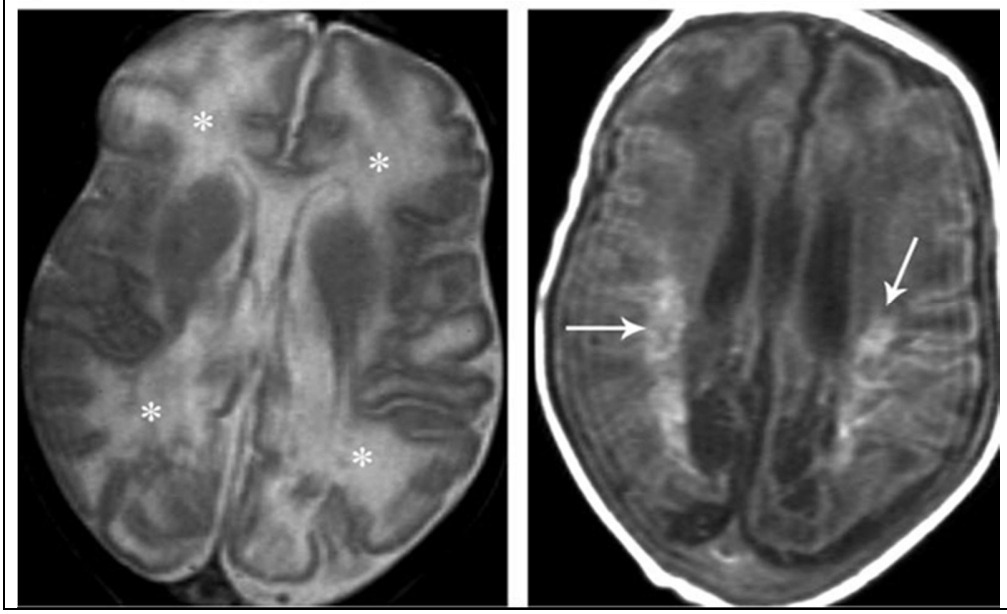
Initial PVL:

- Areas of T1 hyperintensity within larger areas of T2 hyperintensity in the periventricular white matter
- DWI: show early restriction of diffusion with ischemia and coagulative necrosis, findings that normalize within 5–7 days



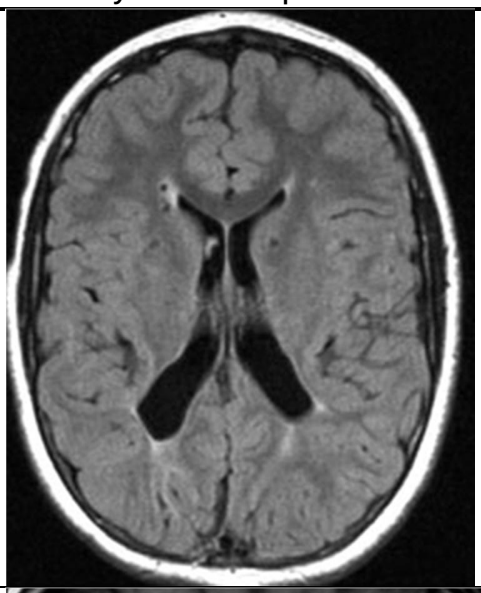
Subsequent PVL

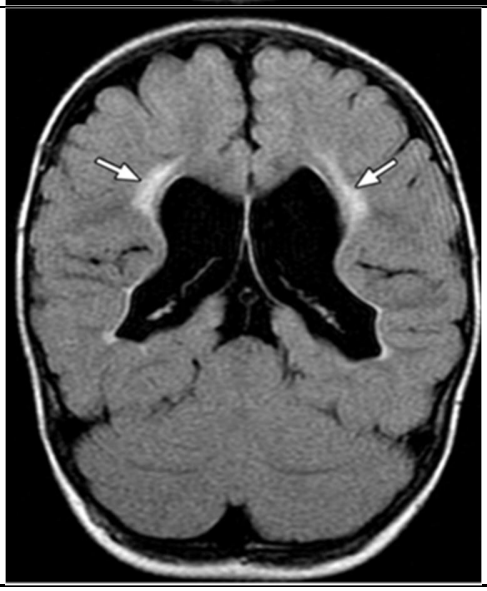
- Cavitation and periventricular cyst formation, features that are required for a definitive diagnosis of PVL, develop 2–6 weeks after injury.

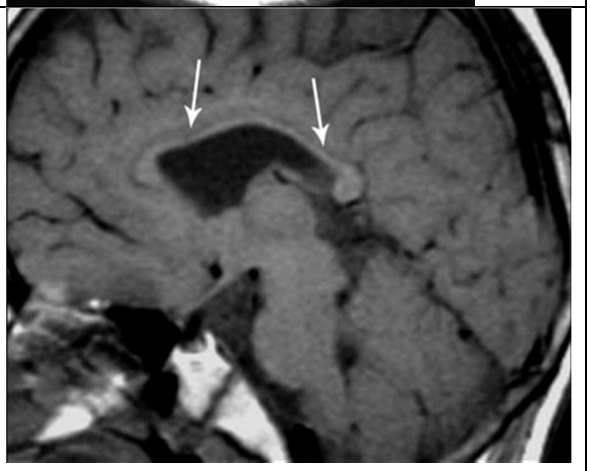


End-stage PVL

- Increased SI and a few tiny cysts in the periventricular white matter
- Decrease in volume of the adjacent periventricular white matter
- Enlargement of the atria of the lateral ventricles
- The walls of the lateral ventricles have a wavy appearance.
- Midsagittal T1 shows thinning of the body of the corpus callosum.







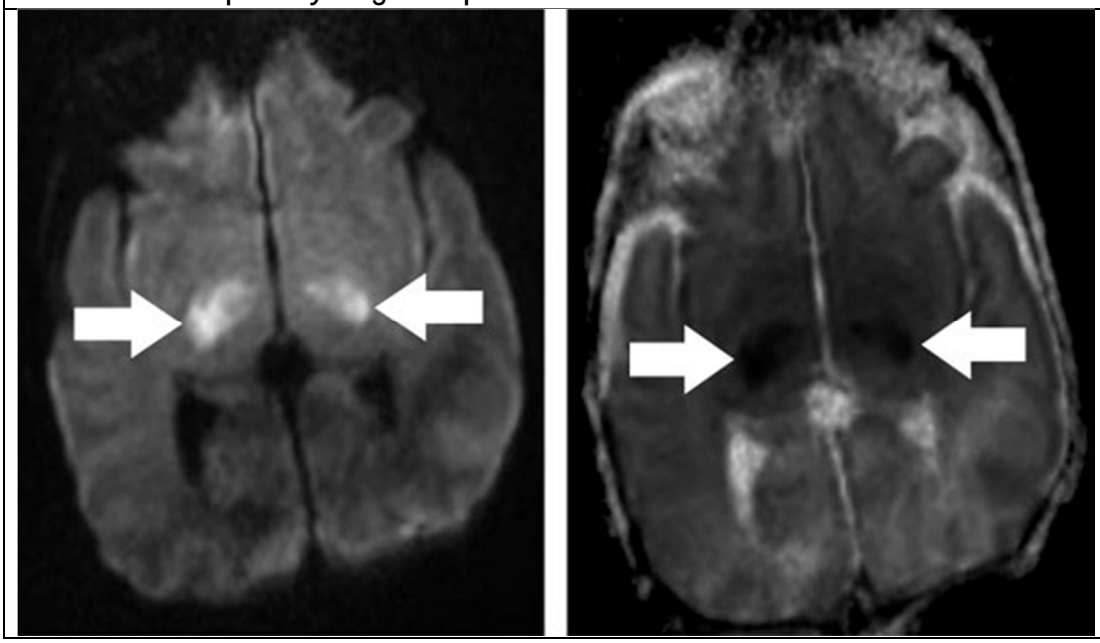
II- Severe Asphyxia in Preterm Infants

Injury to:

- Thalami,
- Dorsal brainstem,
- Anterior part of the cerebellar vermis
- Hippocampi,
- Cerebellum,
- Corticospinal tracts

❖ Imaging features:

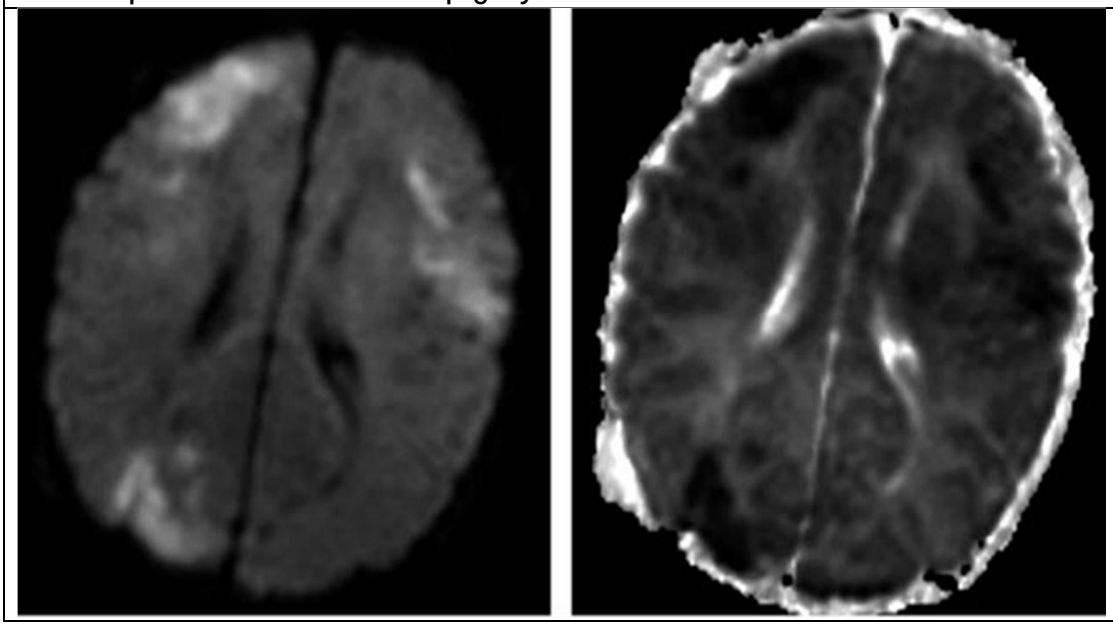
- US: increased echogenicity in the thalami by 48–72 hours following an insult but may also be normal, particularly in the first 2 days.
- CT: hypoattenuation
- T2WI: variable T2 signal
- DWI: Restricted diffusion
- ADC: Low signal
- DWI abnormalities are usually evident in the thalami within 24 hours
- DWI abnormalities are most apparent around days 3–5 following insult and subsequently begin to pseudonormalize



III- Mild to Moderate Asphyxia in Term Infants

1) Parasagittal Watershed Zones Infarctions

- Cortex and the underlying subcortical white matter in
- Parasagittal Watershed Zones:
- NECT: loss of cortical ribbon; WM edema
- T2: cortical swelling with loss of differentiation between gray matter and white matter and hyperintensity
- T1: hypointense
- DWI: Restricted
- MRS: reveals increased lactate concentration in the intervascular zone compared with in the deep gray matter.

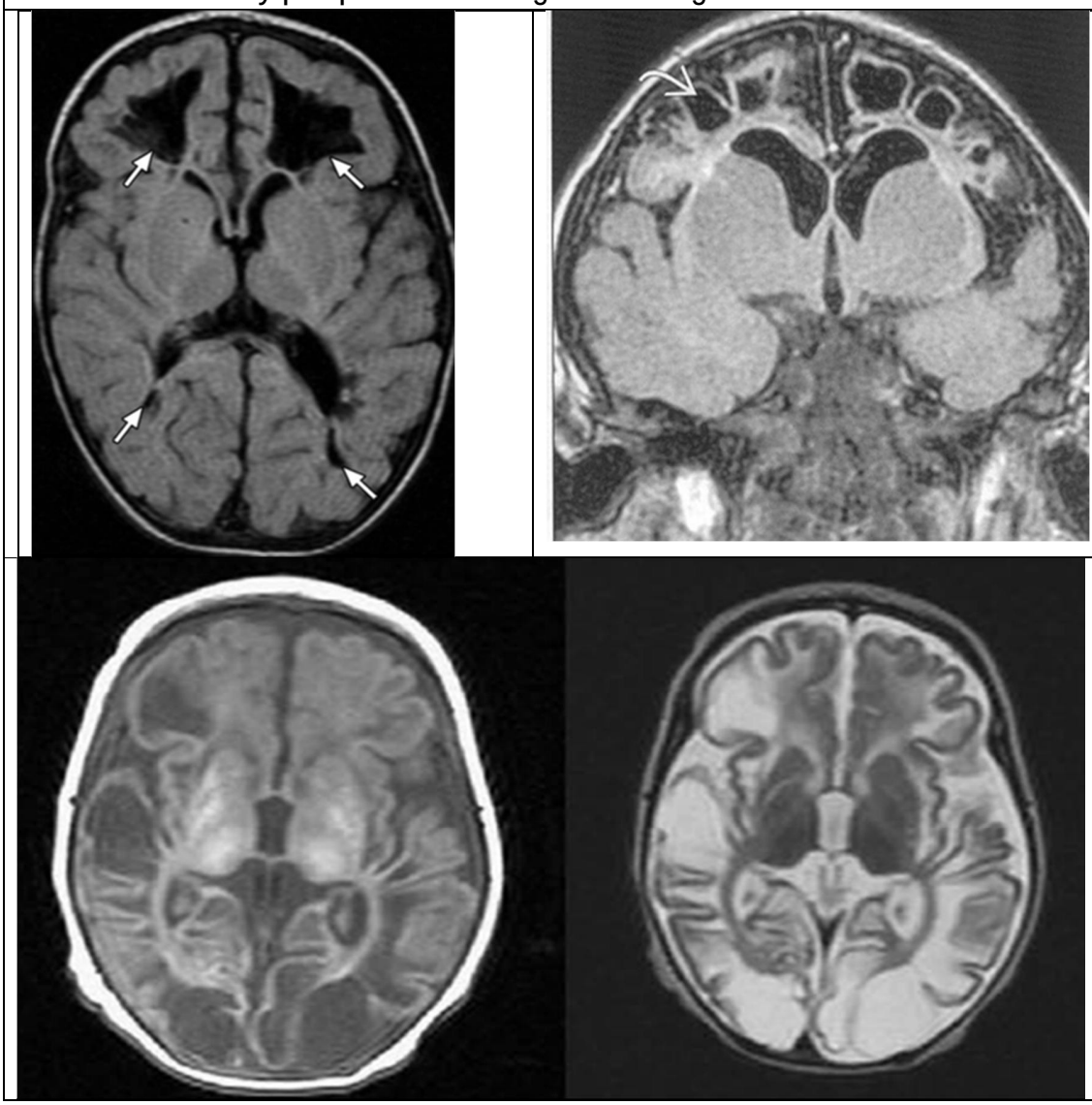


2) Multicystic Encephalomalacia:

- Atrophy with cortical thinning and diminished white matter volume, predominantly in the parasagittal watershed zones.

<u>Imaging findings:</u>

- Cystic encephalomalacia = Low 1+ High T2 + Low FLAIR
- Surrounded by peripheral rim of gliosis of high T2 & FLAIR



IV- Severe Asphyxia in Term Infants

1) Deep GM + Perirolandic cortex

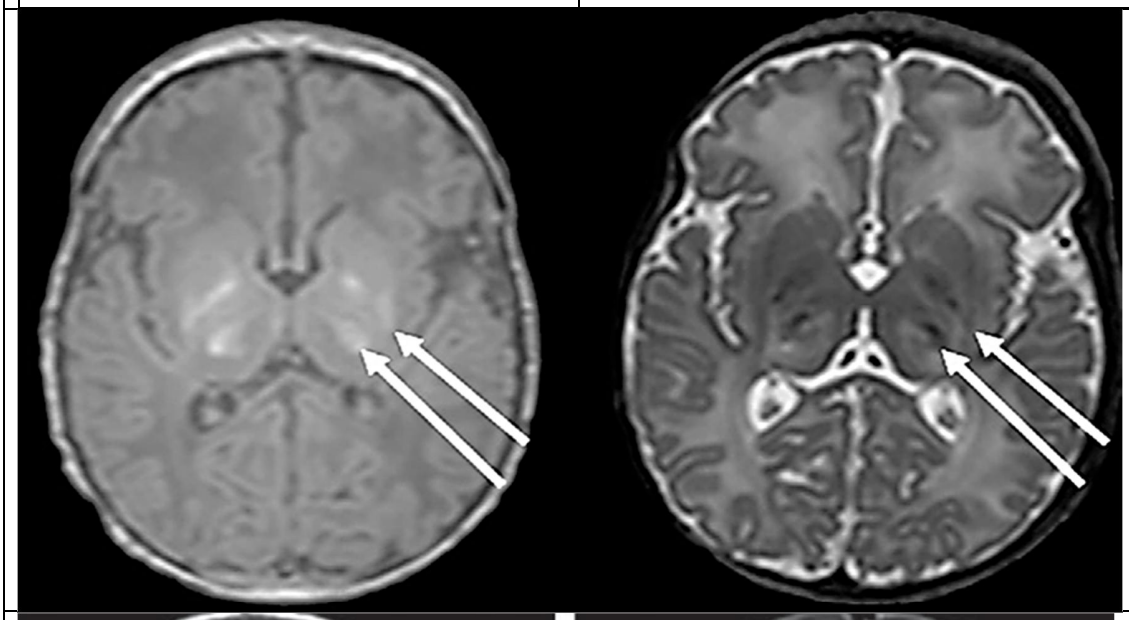
- The metabolically active tissues in the brain of the term neonate are most susceptible to injury in profound hypotension and include:
    - Deep gray matter
        - Ventro-Lateral (Anterolateral) thalami,
        - Globus pallidus
        - Posterior putamina,
        - Hippocampi,
        - Dorsal Brain stem,
        - Lateral geniculate body
    - Sensorimotor cortex (perirolandic cortex).

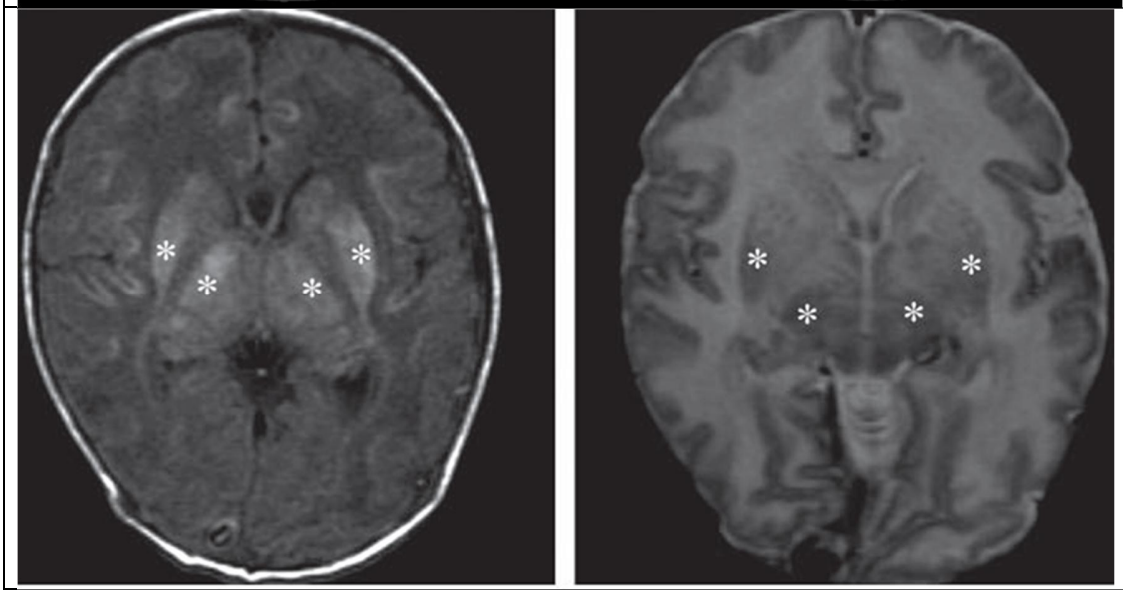
Imaging Findings:

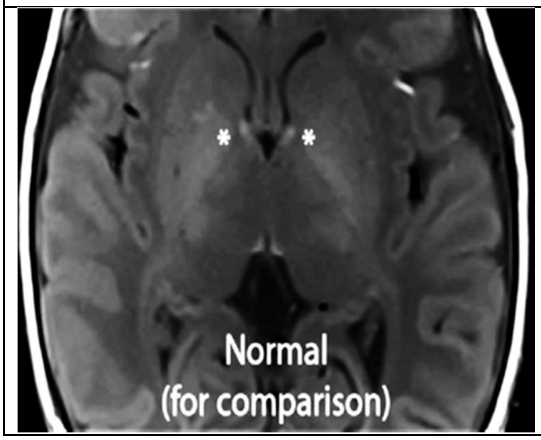
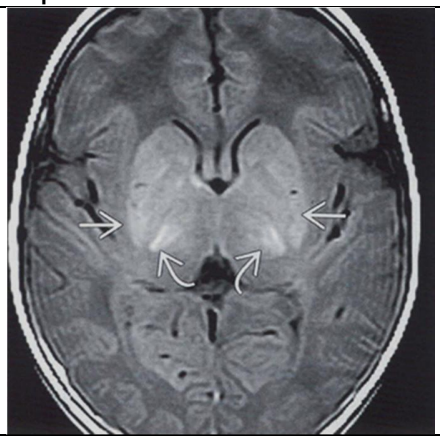
- T1 WI:
    - Low T1 signal of the posterior limb internal capsule = Loss of normal high T1 signal of normally myelinated posterior limb internal capsule
    - High T1 signal of the anterolateral thalamus, posterolateral putamen, globus pallidus and perirolandic cortex.
- T2 WI: Variable signal
- DWI: Restricted diffusion
- ADC: Low signal

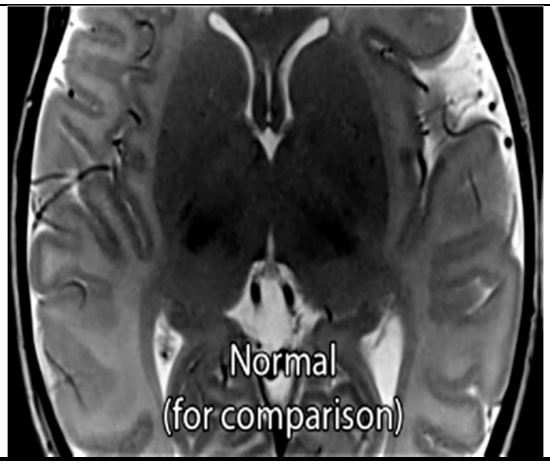
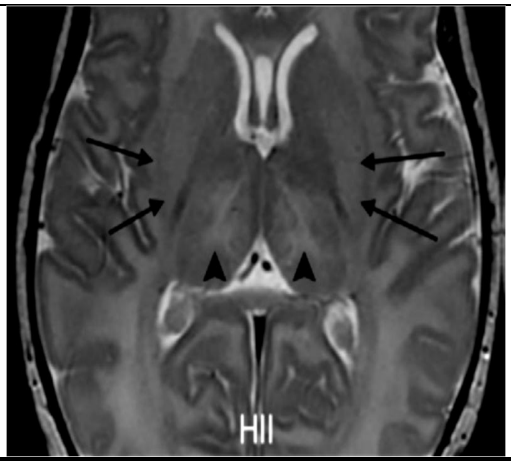
- Anterolateral thalamus + Posterolateral putamen + Globus pallidus
  High T1 + Low T2
- Loss of T1 high signal of posterior limb of internal capsule

Axial T1	Axial T2

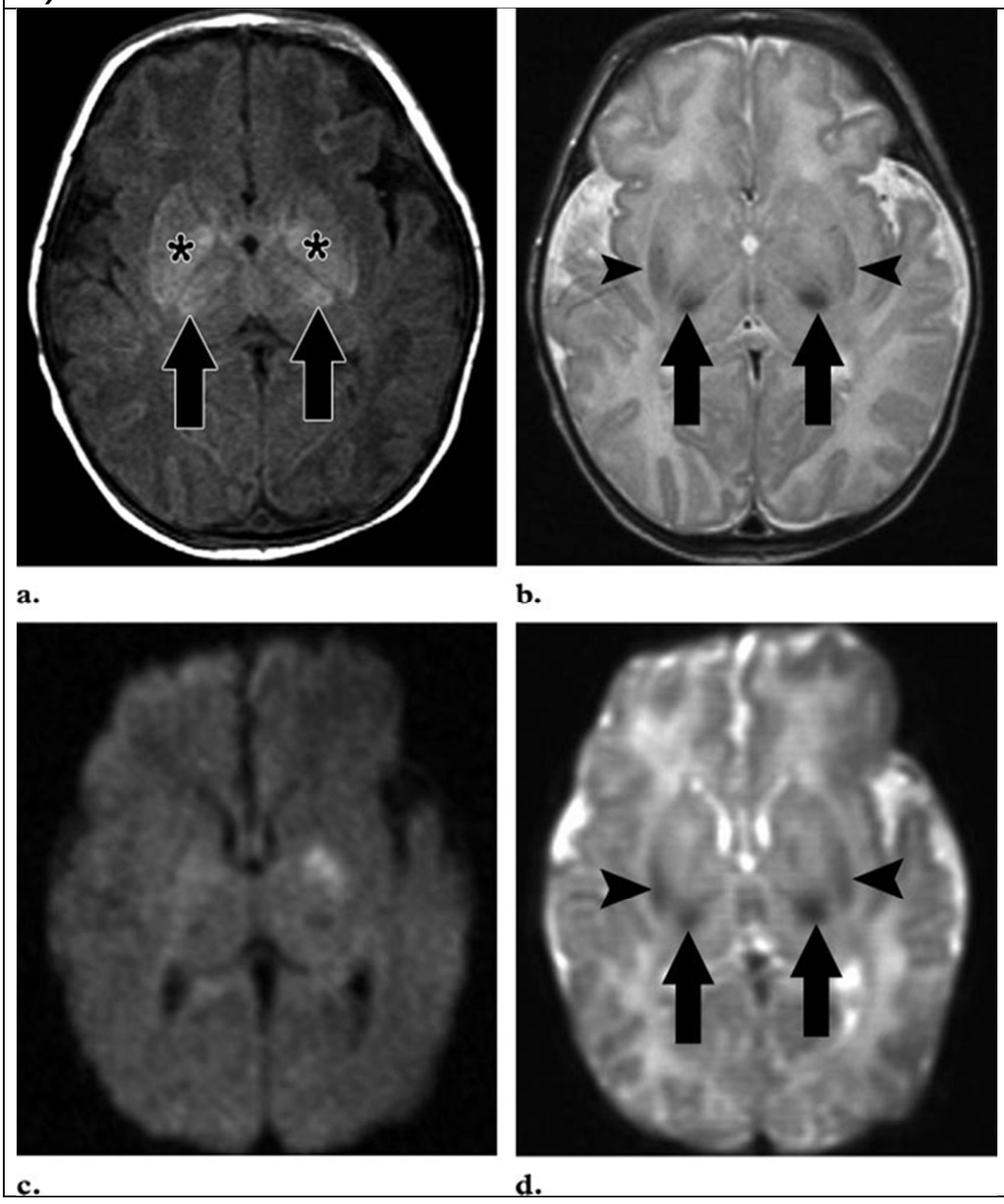




T1 of Normal brain:	T1 of HIE
Globus pallidus (*) is more hyperintense than the putamina.	High T1 signal of anterolateral thalamus + posterolateral putamen + globus pallidus
The posterior limb of the internal capsule + ventrolateral thalami are more hyperintense than the posterolateral putamen.	Low signal of the posterior limb internal capsule = Loss of normal high T1 signal of normally myelinated posterior limb internal capsule
	

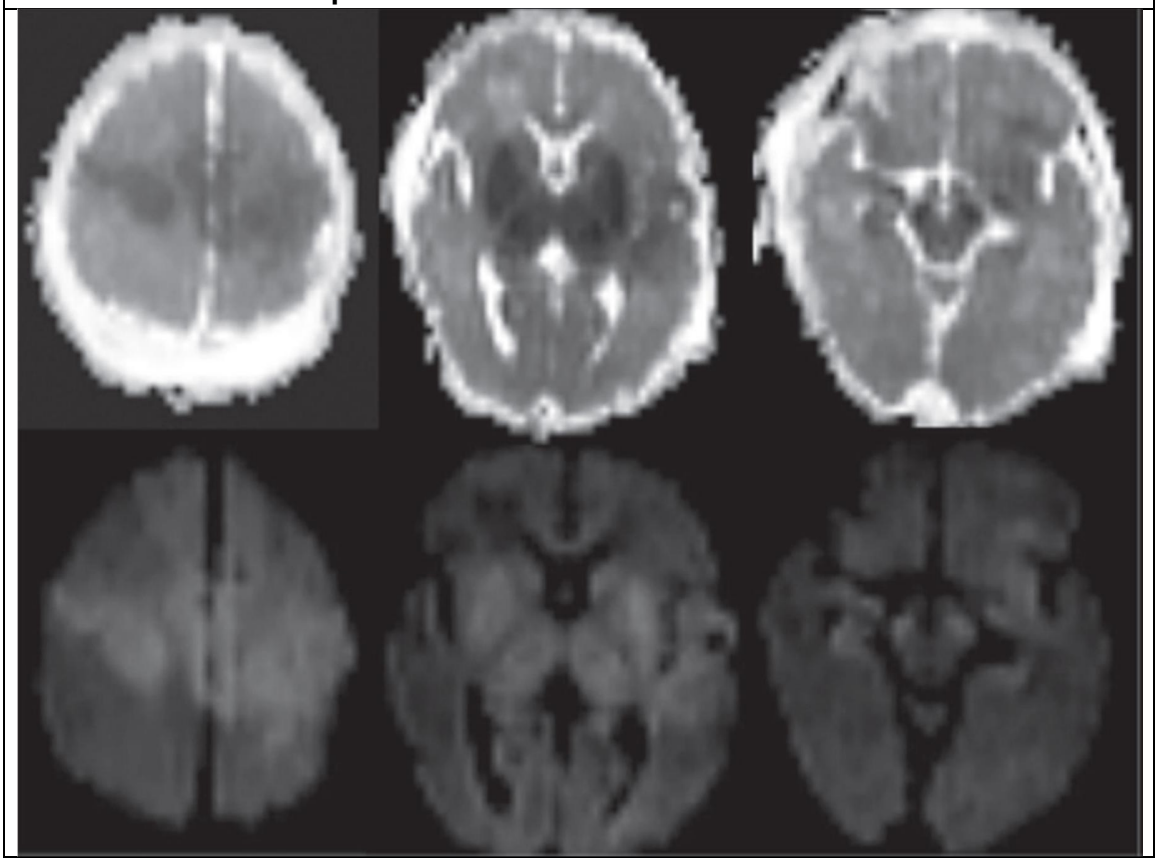
T2 of Normal brain:	T2 of HIE
T2 hypointense of Ventrolateral thalami and posterior limbs of the internal capsule	T2 hperintense signal in the lentiform nuclei, particularly the posterolateral putamina (arrows) + both thalami
	

Lentiform nuclei (*) and ventrolateral thalami (arrows)
A) High T1 signal
B) Low T2 signal
C) Restricted DWI
D) Low ADC



Restricted DWI + Low ADC

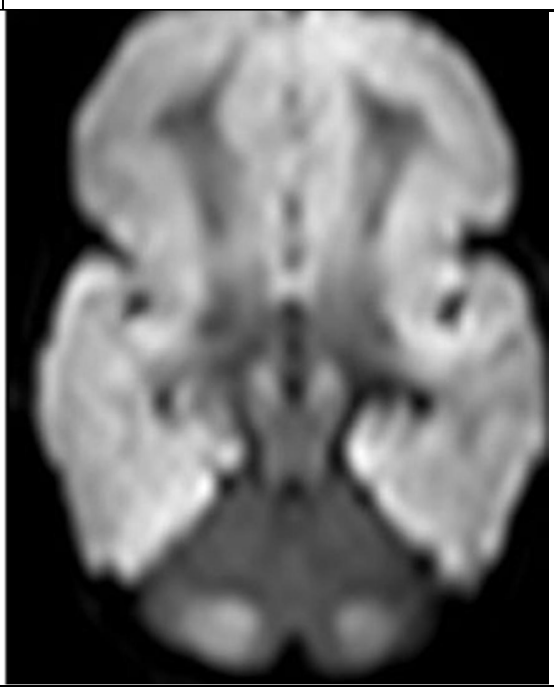
- Basal ganglia
- Bilateral thalami
- Perirolandic cortex
- Mesial temporal lobes.



2) White Cerebrum

- Severe involvement of the subcortical white matter and cortex can be seen with relative sparing of the immediate periventricular white matter and central grey matter, referred to as the 'white cerebrum'.
- This condition tends to be fatal.

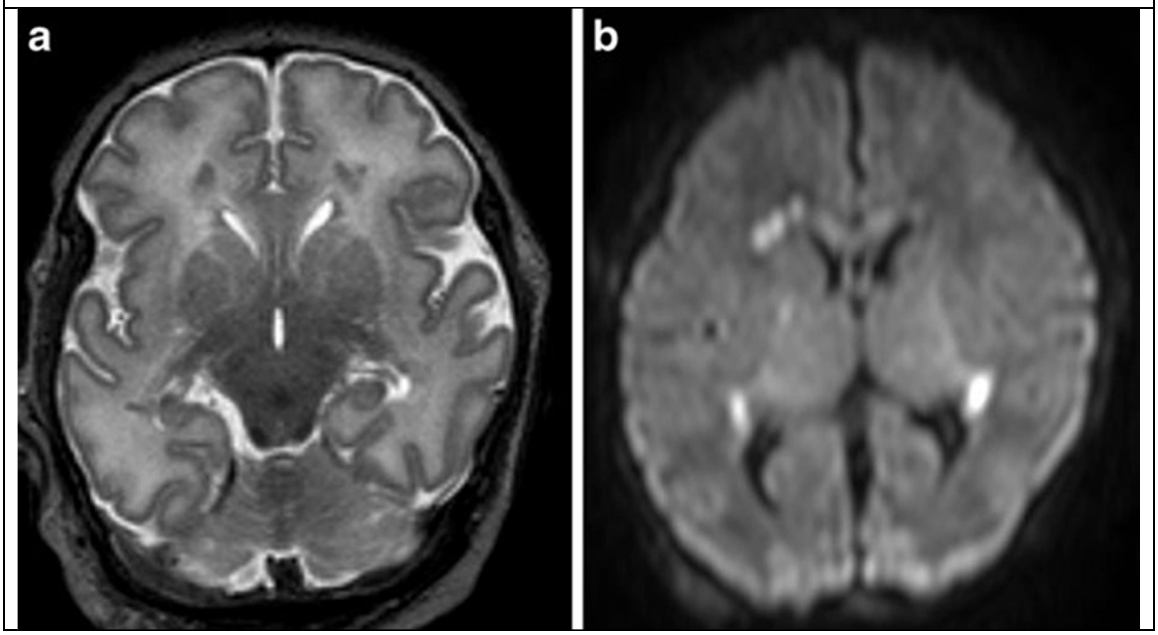
T2	DWI
• Increased signal in the white matter with loss of cortical ribbon. • Relative sparing of the basal ganglia and immediate periventricular white matter	• Restricted diffusion of the cerebral WM • Restricted diffusion of the cerebral peduncles • Striking discrepancy in SI with the cerebellum.



3) Punctate White Matter Lesions

- Another pattern of brain injury consists of “lesions restricted to the periventricular white matter”, not dissimilar from the so-called punctate white matter lesions in the preterm infant.

- T2: low signal intensity punctate white matter lesions
- DWI: Restricted diffusion



II- Intraparenchymal Hemorrhage

❖ CT imaging of hemorrhage

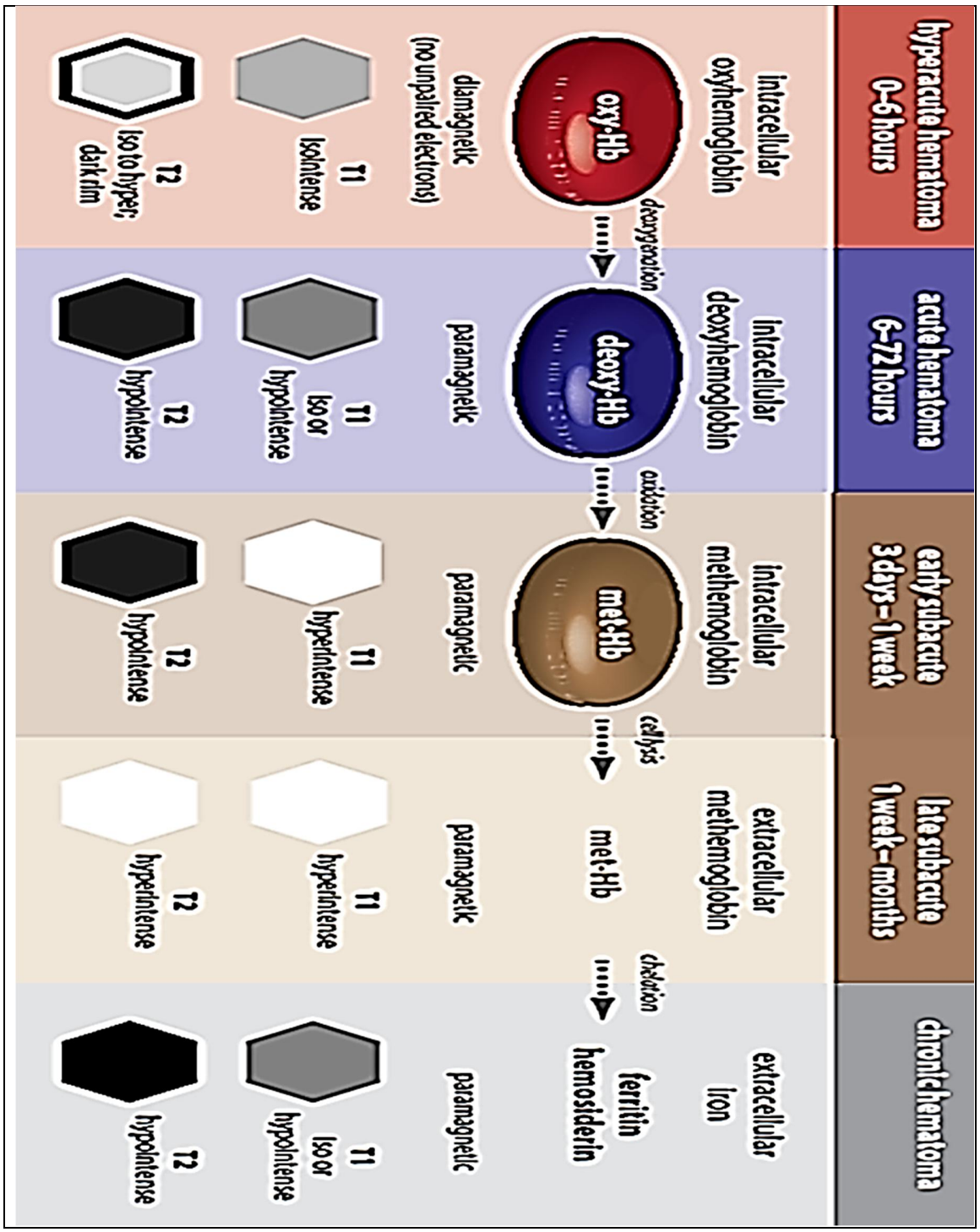
- NCCT is usually the first imaging study performed in the emergency setting for a patient with a sudden neurologic deficit, headache, seizure, or altered level of consciousness.
- CT is highly sensitive for detection of hyperacute/acute intracranial hemorrhage
- NCCT => Hyperattenuating relative to brain parenchyma and CSF.
- CT appearance depends on electron density of clot.
- Isodense with brain:
    - o Rapid hemorrhage
    - o Coagulopathy
    - o Severe anemia (hemoglobin <8 mg/dL).

❖ MR imaging of hemorrhage

- The characteristics of blood products change on T1-and T2- as the iron in hemoglobin evolves through physiologic stages:

→ Intracellular oxyhemoglobin => deoxygenation
→ Intracellular deoxyhemoglobin => oxidation
→ Intracellular methemoglobin => cell lysis
→ Extracellular methemoglobin => chelation
→ Hemosiderin and ferritin

Stage	Time		Hb Stage	NCCT	T1	T2
					Changes occur from peripheral to central	
Hyperacute	< 6hrs	IC	Oxy Hb	Hyper	Iso	Hyper
Acute	6hrs – 3days	IC	Deoxy Hb	Hyper + Hypodense rim	Iso	Hypo
Early Subacute	4-7 days	IC	Met Hb	Iso	Hyper	Hypo
Late subacute	1-4w	EC	Met Hb	Iso ± Enh rim	Hyper	Hyper
Chronic	Months - years	EC	Hemosiderin	Hypo	Hypo	Hyper


hyperacute hematoma
0–6 hours
acute hematoma
6–72 hours
early subacute
3 days–1 week
late subacute
1 week–months
chronic hematoma
Intracellular
oxyhemoglobin
oxy-Hb
diamagnetic
(no unpaired electrons)
T1
isointense
T2
iso to hyper;
dark rim
deoxygenation
Intracellular
deoxyhemoglobin
deoxy-Hb
paramagnetic
T1
iso or
hypointense
T2
hypointense
oxidation
Intracellular
methemoglobin
met-Hb
paramagnetic
T1
hyperintense
T2
hypointense
cell lysis
extracellular
methemoglobin
met-Hb
paramagnetic
T1
hyperintense
T2
hyperintense
chelation
extracellular
iron
ferritin
hemosiderin
paramagnetic
T1
iso or
hypointense
T2
hypointense

A) Hypertensive Intraparenchymal Hemorrhage

- Chronic hypertension is the most common cause of spontaneous adult intraparenchymal hemorrhage

Characteristic locations:

- Basal ganglia (65%)
- Thalamus (25%)
- Cerebellum (10%)

Imaging findings:

- Intraparenchymal hemorrhage with CT and MRI features as described before
- Periventricular white matter disease and prior lacunar infarcts

NCCT:

- High density hematoma in the thalamus
- Surrounded by hypodense rim (Clot retraction)
- May rupture into the adjacent lateral ventricle causing intraventricular hemorrhage and hydrocephalus

B) Cerebral Amyloid Angiopathy (CAA)

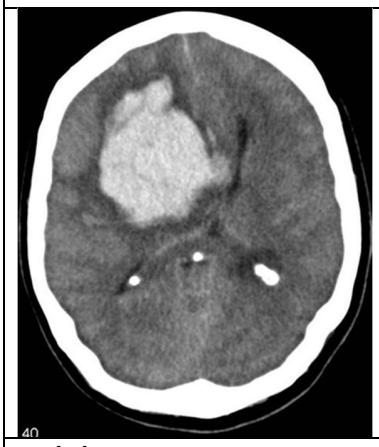
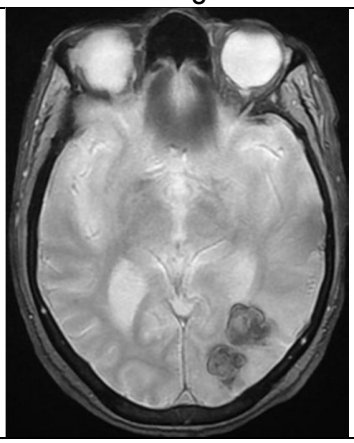
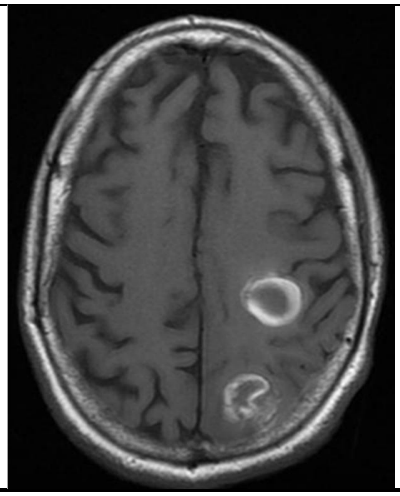
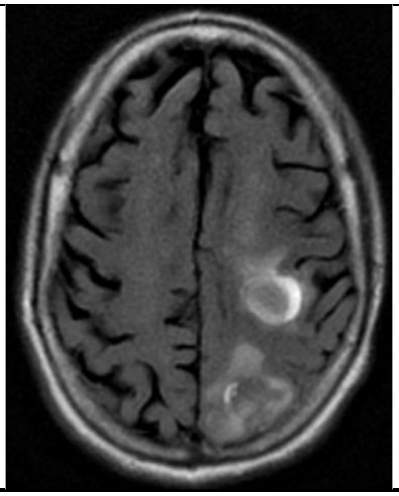
- 2nd most common cause of nontraumatic hemorrhage
- Normotensive elderly patients.
- most often presents clinically as an intracerebral haemorrhage
- It is usually not associated with systemic amyloidosis.

Microhaemorrhages:

- Small focal cortical regions of signal drop out best seen on T2* as regions of blooming

Cerebral (Lobar) Haemorrhage :

- usually superficial cortico-medullary junction

NCCT	T2* Blooming
	
Axial T1	**Axial FLAIR**
	

C) Hemorrhagic Neoplasm

- Incidence: 1% of non-traumatic intra-cerebral hemorrhage

1ry Brain Tumors that commonly Hge:

1) Glioblastoma Multiforme (Most common)
2) Anaplastic astrocytoma
3) Pituitary adenoma

Metastatic Tumors that commonly Hge:

1) Melanoma
2) Choriocarcinoma
3) Renal cell carcinoma
4) Lung
5) Breast

Benign	Malignant
• Well defined	• Bizarre shaped
• Complete hemosiderin rim	• Incomplete rim
• No enhancement	• Enhance
•	• Extensive perilesional edema

III- Subarachnoid Hemorrhage (SAH)

Aetiology

1) Trauma 2) Aneurysm 3) Non-aneurysmal (perimesencepalic) haemorrhage 4) Cerebral AVM 5) Dural AVF	6) Coagulopathy 7) Extension of intraparenchymal hemorrhage 8) Idiopathic

- **Trauma** most common cause of subarachnoid hemorrhage (SAH)
- **Aneurysm** is by far the most common cause of non-traumatic subarachnoid hemorrhage

Locations:

- Basal cisterns
- Sylvian fissures
- Subarachnoid space

Imaging findings:

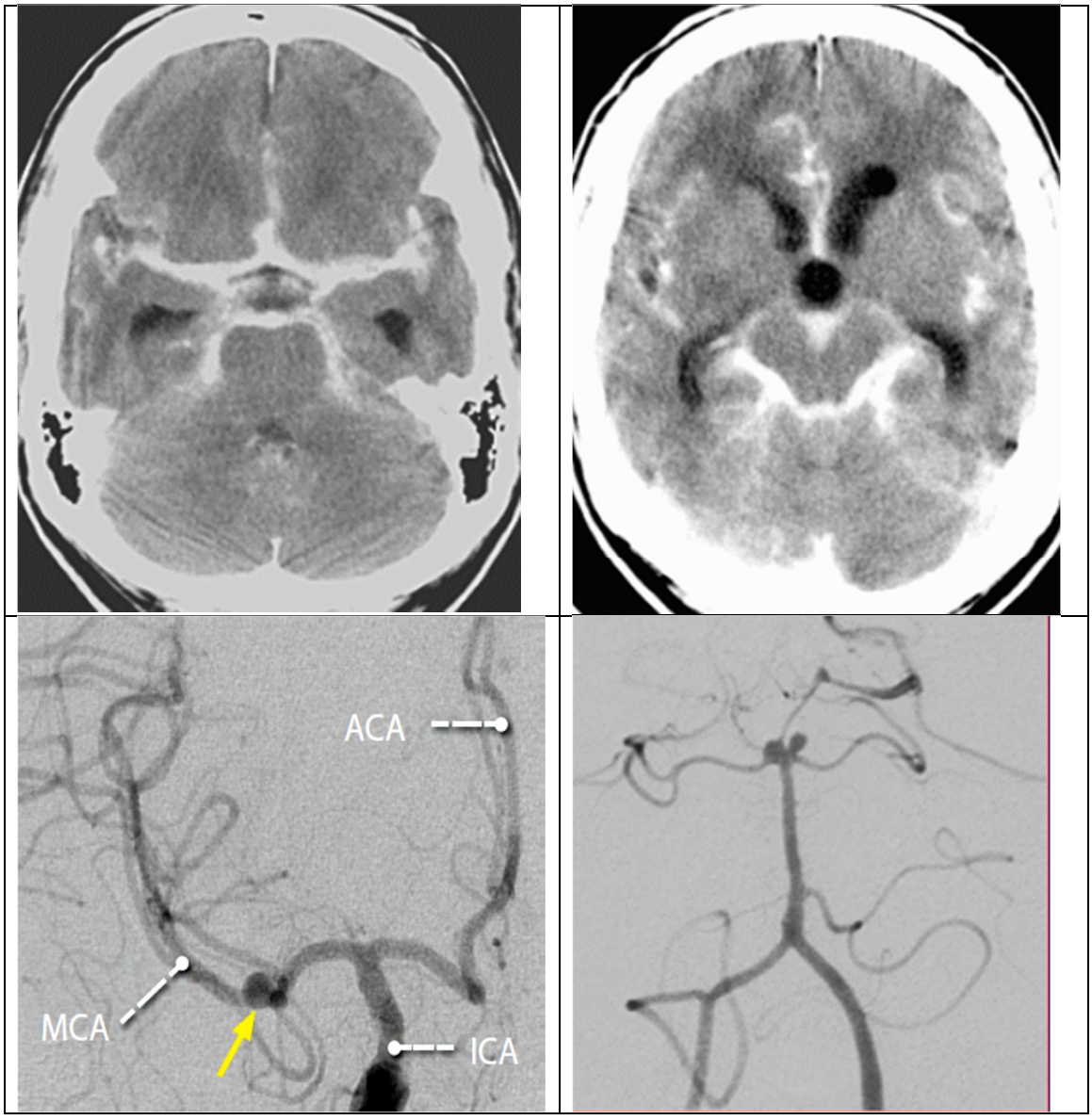
- NCCT >95% sensitive for detecting SAH within the first six hours
- NCCT: hyperdense subarachnoid space
- FLAIR: Hyperintense signal in subarachnoid space
- GRE: Hypointense susceptibility artifact
- +C: No contrast enhancement

DD:

1) SAH
2) Meningitis
3) Leptomeningeal carcinomatosis

❖ Fisher grade classifies the CT appearance of SAH.

- Grade 1 Negative on CT
- Grade 2 SAH <1 mm thick
- Grade 3 SAH >1 mm thick
- Grade 4 Diffuse SAH or intraventricular or parenchymal extension.


ACA
MCA
ICA

❖ Distribution of SAH

- Hemorrhage in the anterior interhemispheric fissure
  ⇨ Acom artery aneurysm (33% of intracranial aneurysms).
- Hemorrhage in the suprasellar cistern
  ⇨ Pcom artery aneurysm (33% of intracranial aneurysms).
- Hemorrhage in the sylvian fissure
  ⇨ MCA aneurysm (20% of intracranial aneurysms).
- Hemorrhage in the perimesencephalic cistern
  ⇨ Basilar tip aneurysm (5% of intracranial aneurysms)
  ⇨ Benign nonaneurysmal perimesencephalic subarachnoid hemorrhage.
- 4th ventricle hemorrhage
  ⇨ PICA aneurysm

❖ Complications of SAH:

i) Acute hydrocephalus

- Rare
- usually obstruction of aqueduct or 4th ventricular outlet by clotted SAH
- Obstructive, non-communicating hydrocephalus
- Asymmetric ventricular dilatation

ii) Delayed hydrocephalus

- Arachnoid granulation defect in CSF resorption
- Obstructive, communicating hydrocephalus
- Symmetric ventricular dilatation

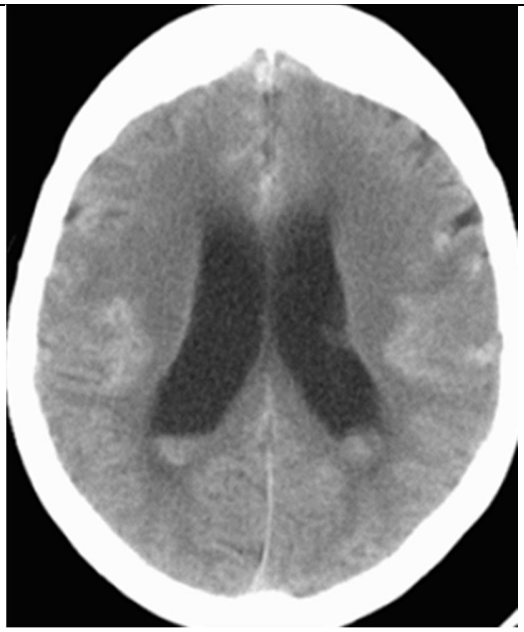
iii) Vasospasm => Ischemia:

- May develop quickly (2-3 days post-injury)
- Peaks 7-10 days after injury
- Threat remains up to 2 weeks
- Uncommon cause of post traumatic infarct.
- Triple H therapy (Haemodilution, Hypertension, Hypervolaemia)

iv) Superficial siderosis

- A condition caused by iron overload of pial membranes due to chronic or repeated subarachnoid bleeding.
- Clinically, patients present with sensorineural deafness and ataxia.
- MRI: iron causes hypointensity on T2 outlining the affected sulci.

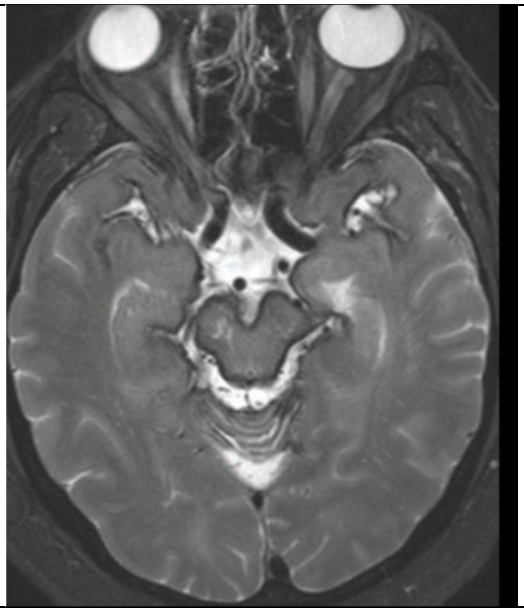
Acute SAH => Hydrocephalus



Superficial siderosis:

<u>T2-weighted images</u>

- Thin hypointense interface at the junction of
- the sulcal surfaces and the subarachnoid space (arrows) of the basal cisterns



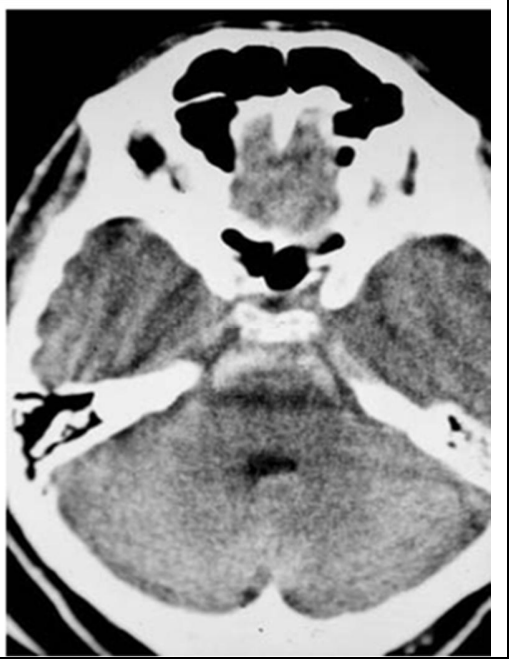
❖ Perimesencephalic SAH

- Nonaneurysmal SAH that is a diagnosis of exclusion
- Better prognosis than hemorrhage due to a ruptured aneurysm.

- Catheter angiography twice, one week apart.
- Both angiograms must be negative.

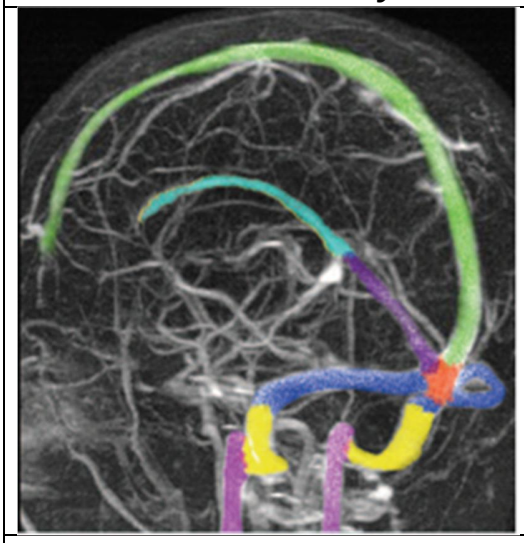
Location:

- SAH must be limited to the perimesencephalic cisterns:
  - Interpeduncular cistern
  - Prepontine cistern
  - CPA cistern
  - Ambient cistern
- The membrane of Liliequist separates the perimesencephalic cistern from the suprasellar cistern and explains why there is no extension of blood superiorly.



IV- CEREBRAL VENOUS DISEASE

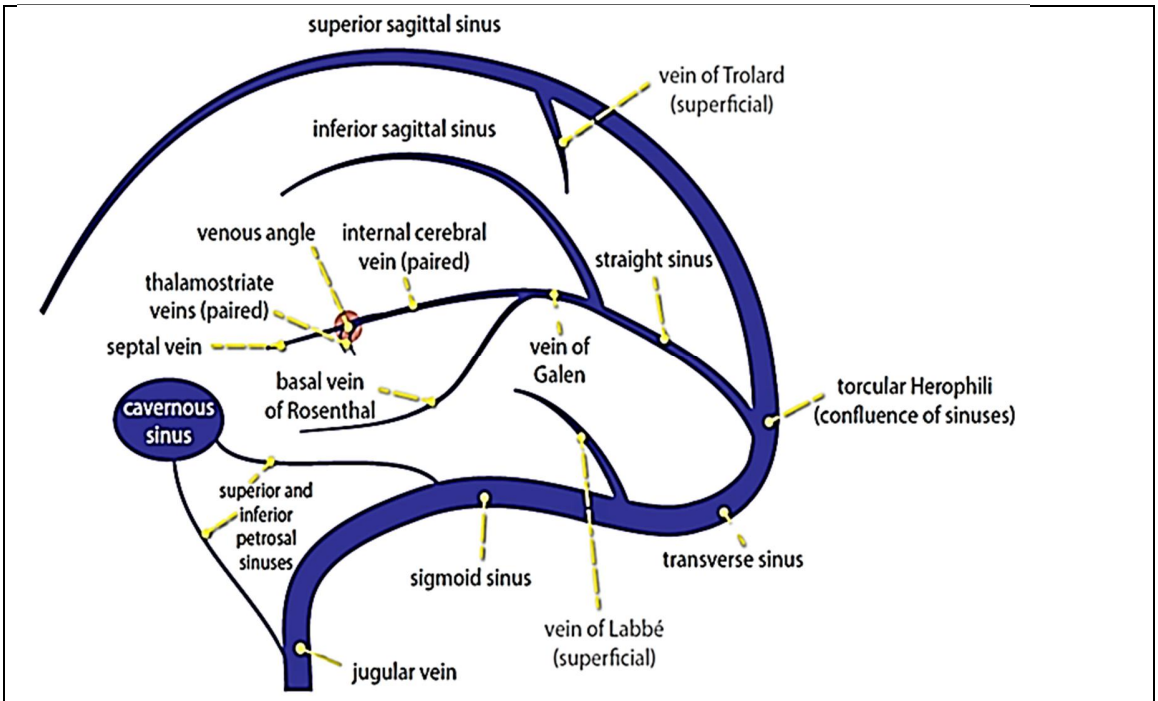
❖ Normal Anatomy of Dural Venous Sinuses:



- **Superior Sagittal Sinus**
- **Inferior Sagittal Sinus**
- **Straight Sinus**
- **Confluence of Sinuses**
- **Transverse Sinuses**
- **Sigmoid Sinuses**
- **IJV And Bulbs**

❖ Normal Anatomy of Deep Venous System:

1= Thalamostriate Vein
2= Septal Vein
3= Internal Cerebral Vein
4= Basal Vein of Rosenthal
5= Vein Of Galen



A) Dural Sinuses

- The superior sagittal sinus (and its tributaries) drains the motor and sensory strips.
- The paired transverse sinuses are usually asymmetric, with the left transverse sinus often hypoplastic.
- The sigmoid sinus connects to the jugular bulb.
- The torcular Herophili is the confluence of the superior sagittal sinus, the transverse sinus, and the straight sinus.

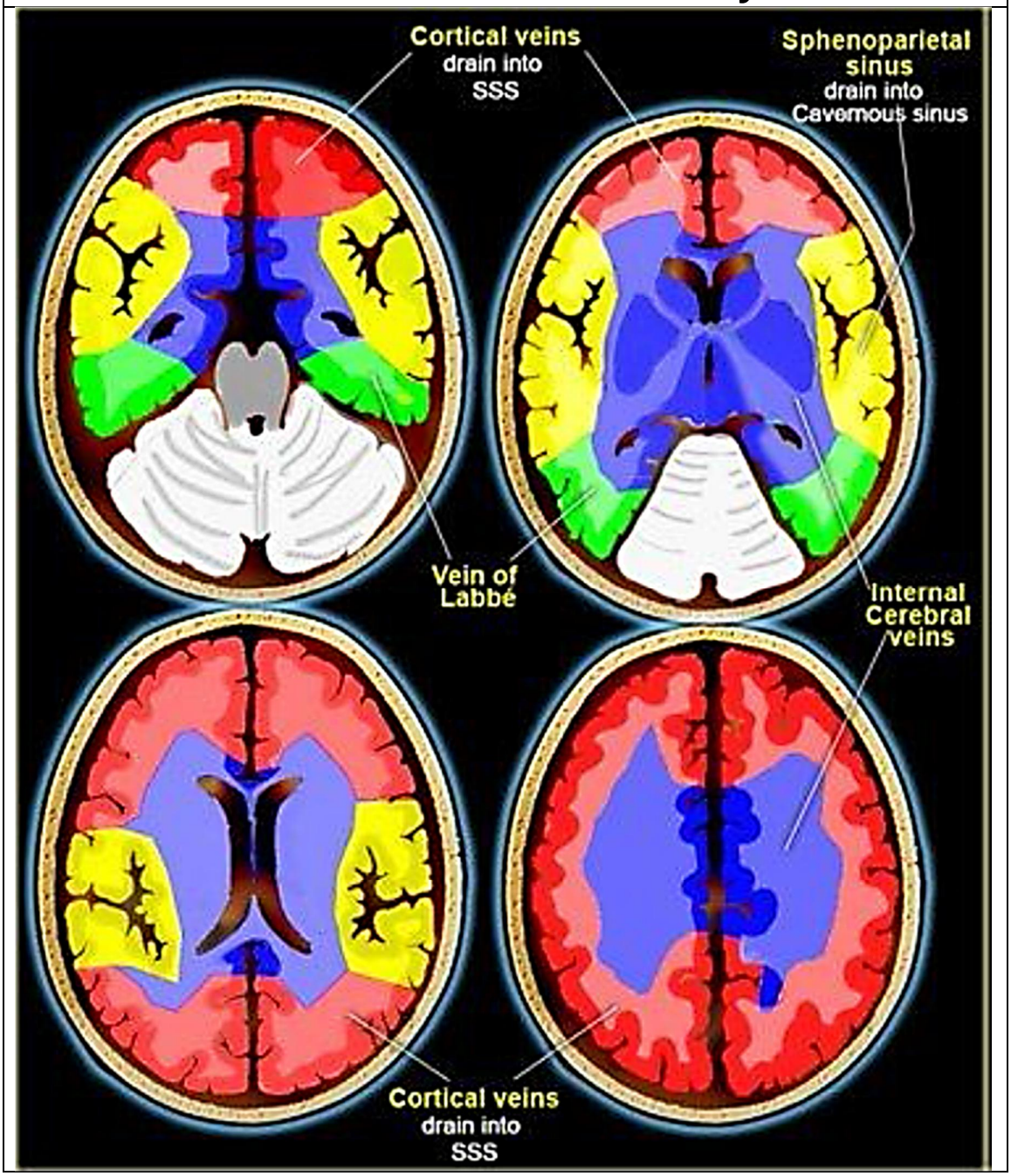
B) Deep Cerebral Veins

- Paired internal cerebral veins, the basal vein of Rosenthal, and the vein of Galen.
- The venous angle (red dot in the diagram above) is the intersection of the septal vein and the thalamostriate veins.
- The venous angle is the angiographic landmark for the foramen of Monro.

C) Superficial Cerebral Veins

- The vein of Trolard connects superficial cortical veins to SSS.
- The vein of Labbé drains the temporal convexity into the transverse or sigmoid sinus.

❖ Cerebral Venous Territory



Cerebral Venous Thrombosis

- Cerebral venous thrombosis results from occlusion of a venous sinus and/or cortical vein and usually is caused by thrombus

❖ Predisposing Factors:

Common	Uncommon
1) Pregnancy - Postpartum 2) Dehydration 3) Drugs: OCPs 4) Coagulopathy 5) Infection: Mastoiditis – Sinusitis 6) Trauma 7) Malignancy: ALL	8) IBD: Crohn's – UC 9) Vasculitis: Behçet 10) SLE 11) Antiphospholipid syndrome 12) Paraneoplastic syndrome

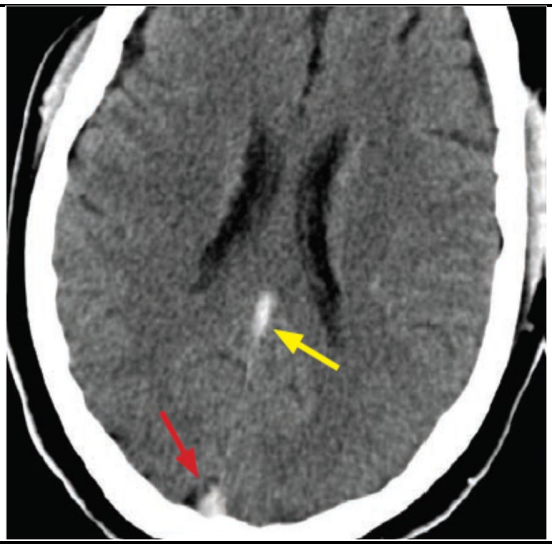
❖ Radiological Appearance of Venous Thrombosis:

<u>NCCT:</u>

- Hyperdense venous sinus
- Hyperdense falx

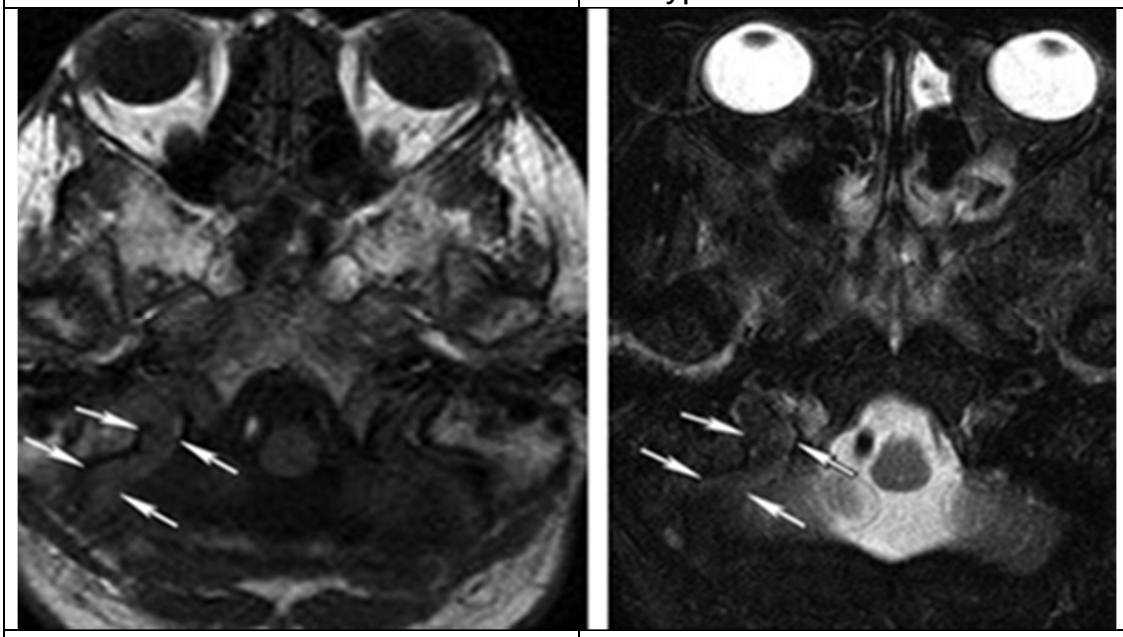
<u>CECT:</u>

- Empty delta sign = Non-enhancing clot surrounded by enhancing dura

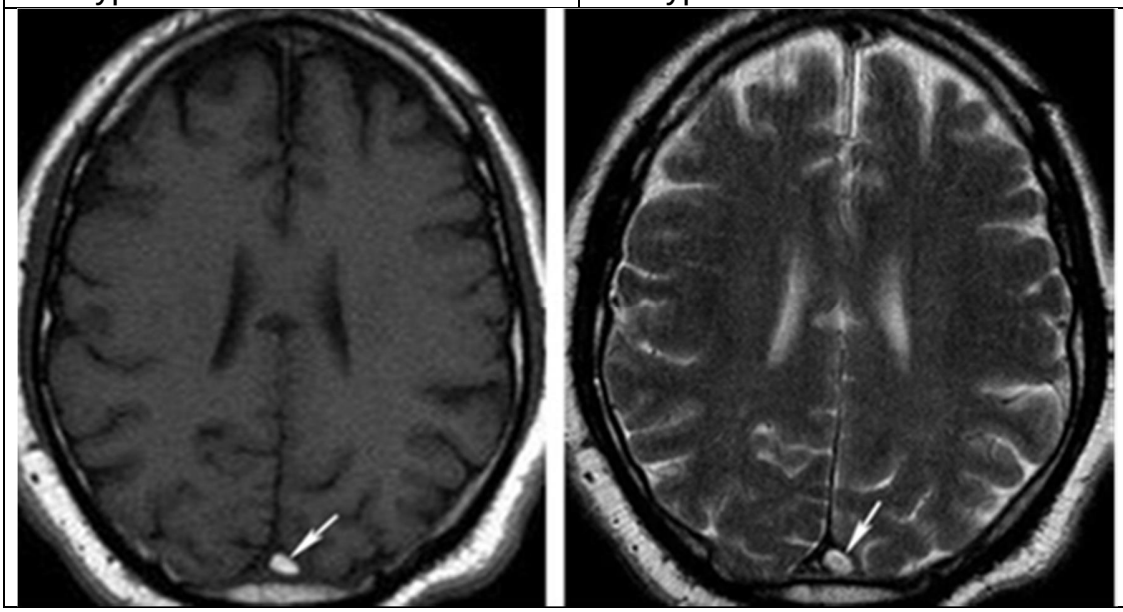


❖ Conventional MRI

❖ Acute thrombus	
T1: Iso	T2: hypo



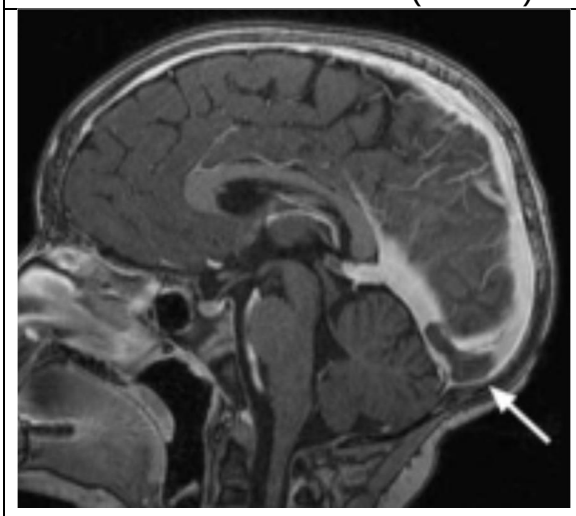
❖ Subacute thrombus	
T1: Hyper	T2: hyper



❖ Contrast Enhanced MRV:

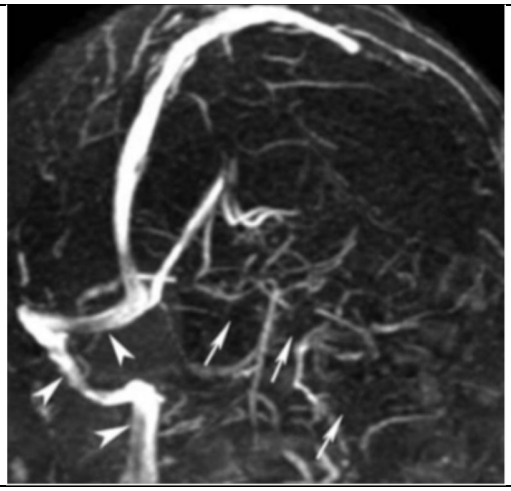
"Empty delta" sign

A nonenhanced thrombus (arrows) surrounded by enhanced sinus walls



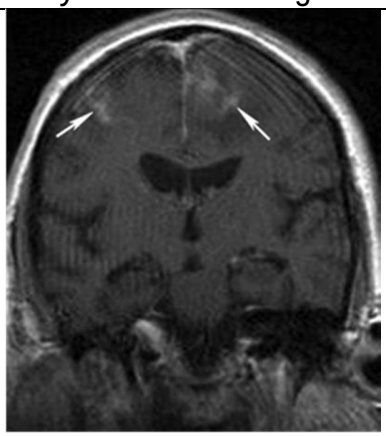
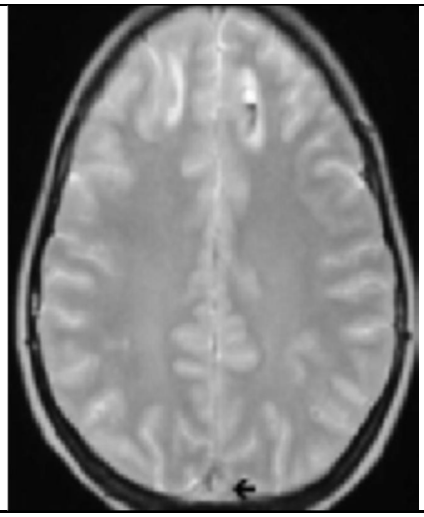
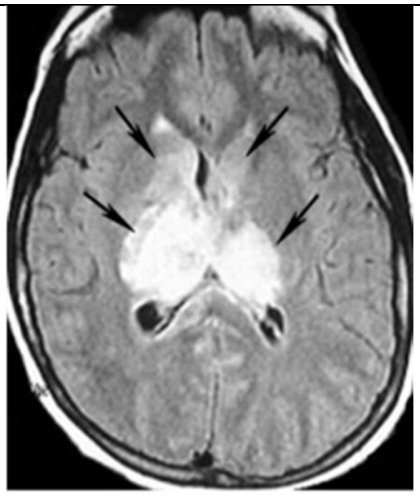
❖ TOF MRV

- Absence of flow signal in the left transverse sinus, left sigmoid sinus, and left internal jugular vein (arrows)
- Normal flow signal in the SSS, right transverse sinus, right sigmoid sinus, and right internal jugular vein (arrowheads).

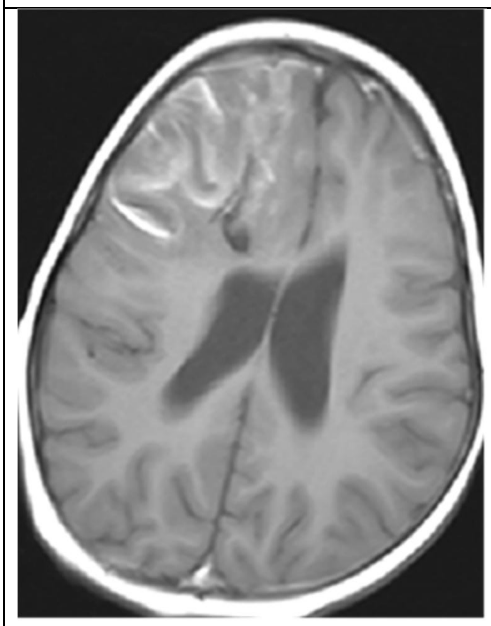
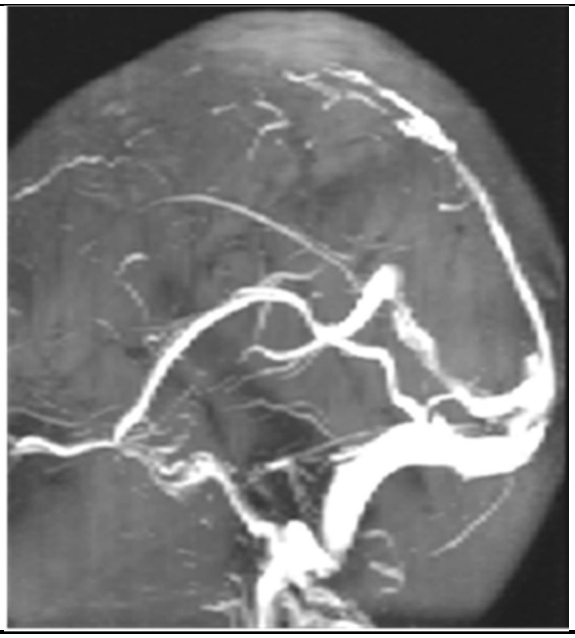


❖ Parenchymal Abnormalities

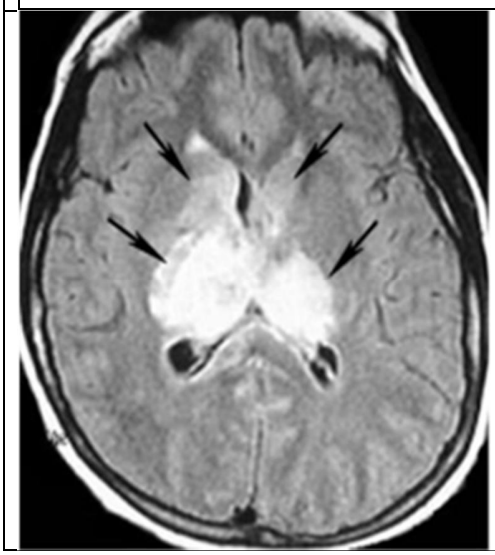
Superior sagittal sinus thrombosis => Infarction of parasagittal high convexity cortex.
Deep venous system thrombosis => Infarction of bilateral thalami.
Transverse sinus thrombosis => Infarction of posterior temporal lobe.

Parenchymal edema: may be secondary to cytotoxic or vasogenic edema.	**Gyral enhancement:** indicates disruption of the blood-brain barrier, may be seen in areas of cytotoxic or vasogenic edema
	
B) Parenchymal hemorrhage: Flame-shaped cortical and subcortical hemorrhagic foci in the parasagittal frontal and parietal lobes are typical findings with SSS thrombosis	Bilateral thalamic hyperintense signal of edema in Thrombosis of the internal cerebral veins, vein of Galen, and straight sinus
	

❖Dural Sinus Thrombosis

T1: Frontal hemorrhagic venous infarction	MRV: SSS thrombosis
	

❖ Deep Venous Occlusion

FLAIR: Characteristic Bilateral thalamic hyperintense signal of edema	MRV: Thrombosis of the internal cerebral veins, vein of Galen, and straight sinus.
	

Cerebral Vascular Malformations

High Flow	Low Flow
• Aneurysm • Cerebral Arteriovenous Malformation (AVM) • Pial AVF • Dural AVF	• Cavernous malformation (cavernoma) • Developmental venous anomaly (DVA) (venous angioma) • Capillary telangiectasia

Cerebral Aneurysms

❖ Aetiology

- Acquired (Degenerative): Atherosclerosis + Hypertension (90-95%)
- Idiopathic
- Congenital
- Vasculitis
- Fibromuscular dysplasia
- Infection = Mycotic aneurysm
- Trauma / Iatrogenic
- Tumor = Oncotic aneurysm

❖ Associations:

- Ehlers-Danlos syndrome (type IV)
- Marfan syndrome.
- Ehlers-Danlos syndrome
- Autosomal dominant polycystic kidney disease (ADPKD)
- Coarctation of aorta
- Bicuspid aortic valve
- Neurofibromatosis type 1 (NF1)
- Hereditary haemorrhagic telangiectasia
- Alpha 1 antitrypsin deficiency
- Cerebral arteriovenous malformation : flow related aneurysm
- Fibromuscular dysplasia

❖ Classification according to Location:

- 90-95% arise from Circle Of Willis
  - Anterior circulation: 90%
    - ACA / Acom complex: ~30 - 40%
    - ICA / Pcom junction: ~30%
    - MCA (M1/M2 junction) bifurcation: ~ 20 - 30%
  - Posterior circulation: 10%
    - Basilar tip
    - SCA
    - PICA
- 5% distal to COW (often traumatic, mycotic, oncotic)

❖ Classification according to Size:

- Small (< 1 cm)
- Medium (1 - 2.5 cm)
- Giant (> 2.5 cm)

❖ Classification according to Shape:

- Saccular = Berry
- Fusiform

❖ Classification according to Type:

- True = contain 3 histologic layers of the vessel wall
- False = Pseudoanerysm

❖ Classification according to Number:

- Solitary = 70 – 80%
- Multiple = 20 – 30%

❖ Imaging Findings:

❖ Angiography

- Identify aneurysm = contrast filled outpouching
- No – Site – Size – Shape
- Relationship to parent vessel = Neck
- Collateral circulation
- Detect complications

<u>Causes Of Negative Angiogram</u>

⇨ <u>Repeat Angiography will be +ve in 10 – 20 %:</u>

- Incomplete study
- Ruptured microaneurysm
- Thrombosed aneurysm
- SAH may be non-aneuryms:
  - Occult vascular malformation
  - Neoplasm
  - Non-=aneurysmal perimesencephalic hemorrhage

❖ CT Angiography

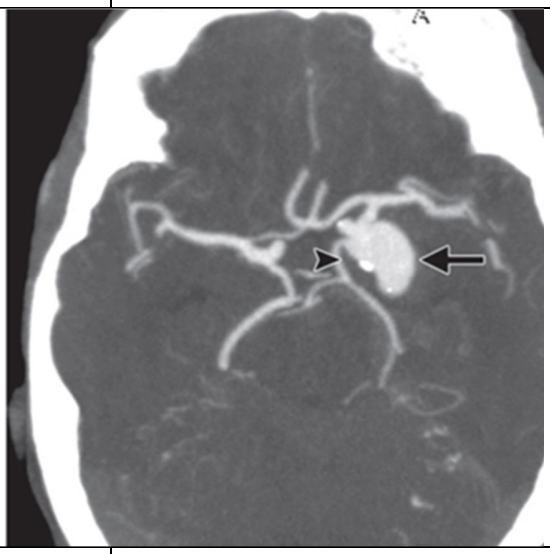
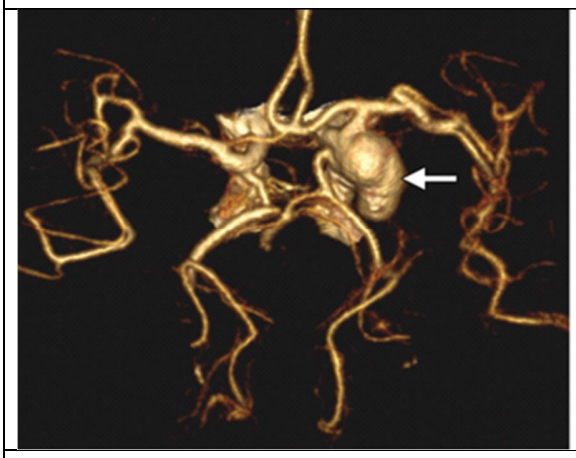
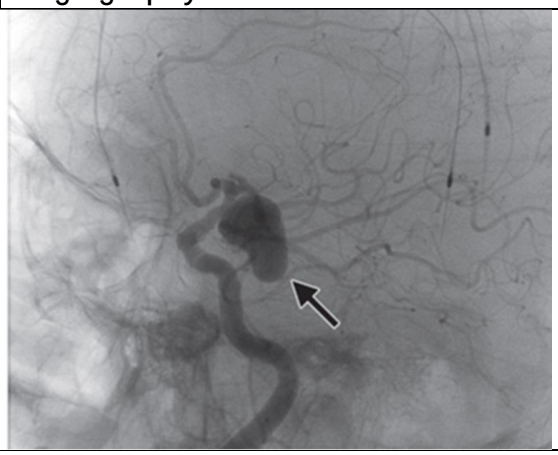
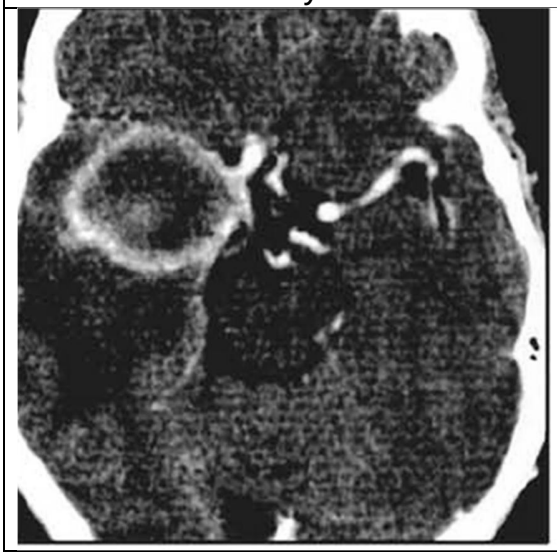
- Aneurysm = Hyperdense intense uniform enhancing outpouching
- ± peripheral curvilineal calcification
- ± peripheral hypodense thrombus

❖ MRI

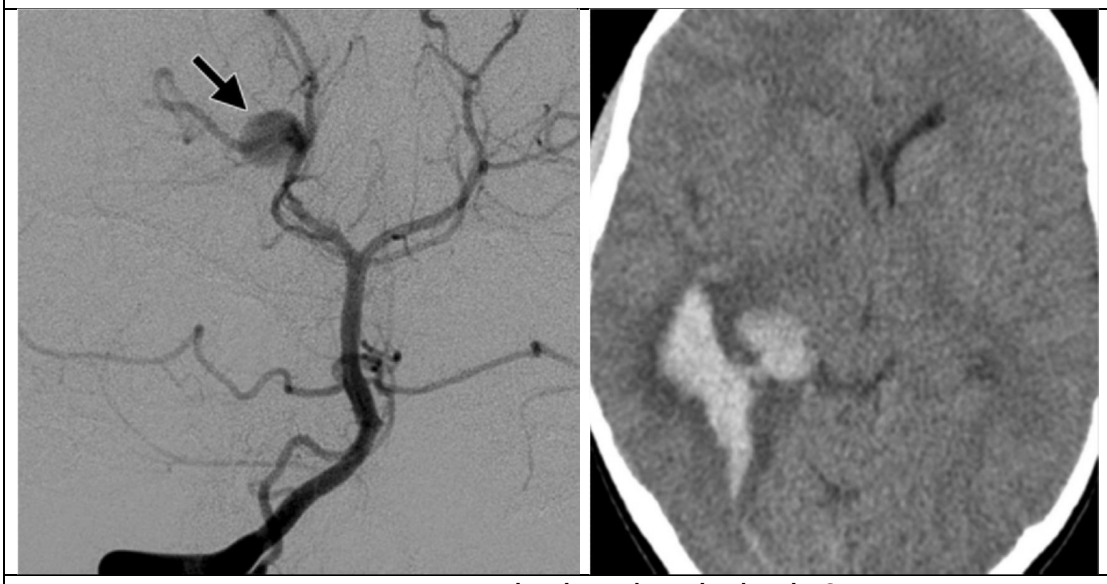
- Patent aneurysm:
  - Rapid flow => Flow void
  - Slow or turbulent flow => Isointense + Misregistration artifact
  - Pulsations => Phase encoding artifact
- Partially thrombosed aneurysm:
  - Thrombus => Multilayered laminated signal in the wall
  - Patent lumen => Flow void

❖ MRA

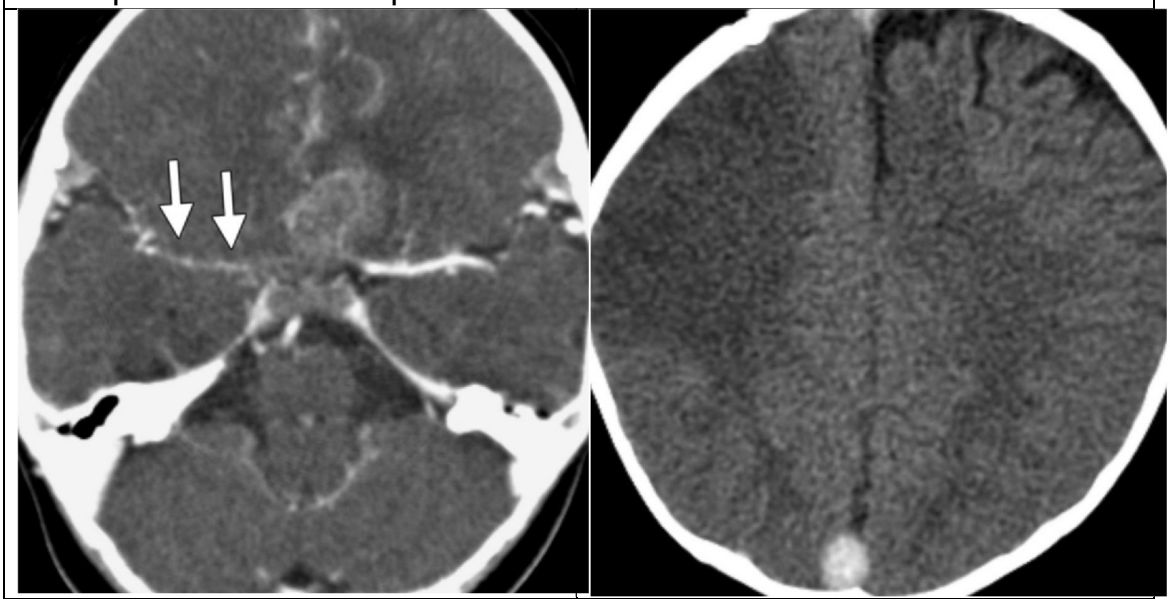
- Non-invasive screening for aneurysms in patients with familial risk.
- 87% sensitive for aneurysms > 5mm

NCCT: SAH	CTA MIP
	
3D CTA	**Angiography**
	
Thrombosed Aneurysm	
	

Rt PCA aneurysm => Rt temporal intaraparenchymal hge + Rt ambient cistern SAH



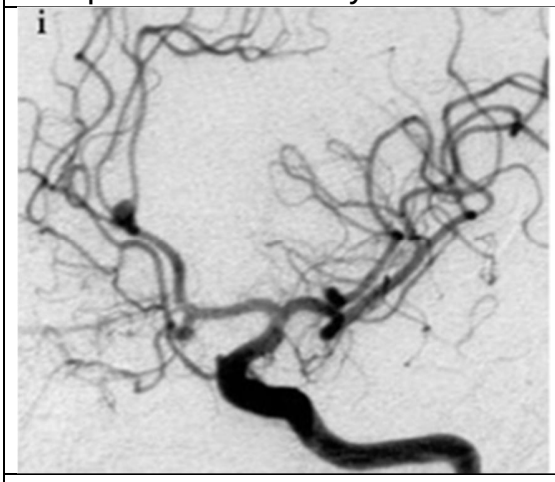
Acom aneurysm rupture => Anterior interhemispheric SAH +
MCA spacm => Rt fronto-parietal infarction



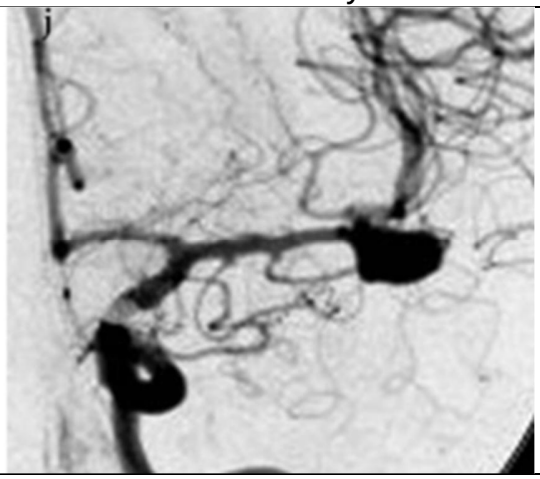
❖ Saccular = Berry Aneurysms

- Focal outpouching of the arterial wall
- Location Most commonly arising at a branch point in the circle of Willis
- Saccular aneurysms are caused by a combination of hemodynamic stress and acquired degeneration of the vessel wall.

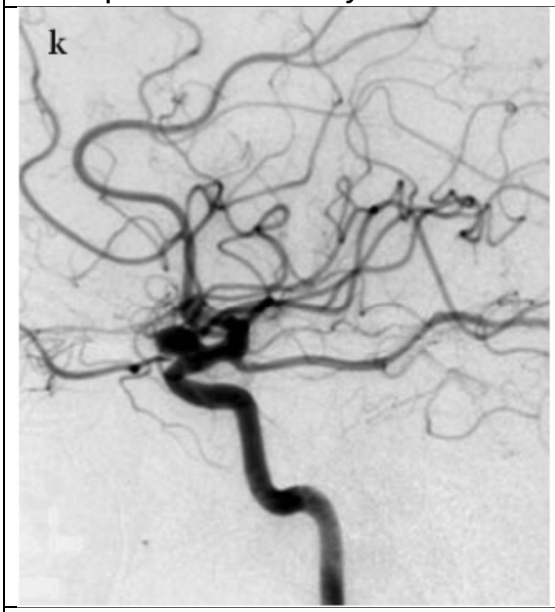
ACA pericallosal aneurysm



MCA bifurcation aneurysm

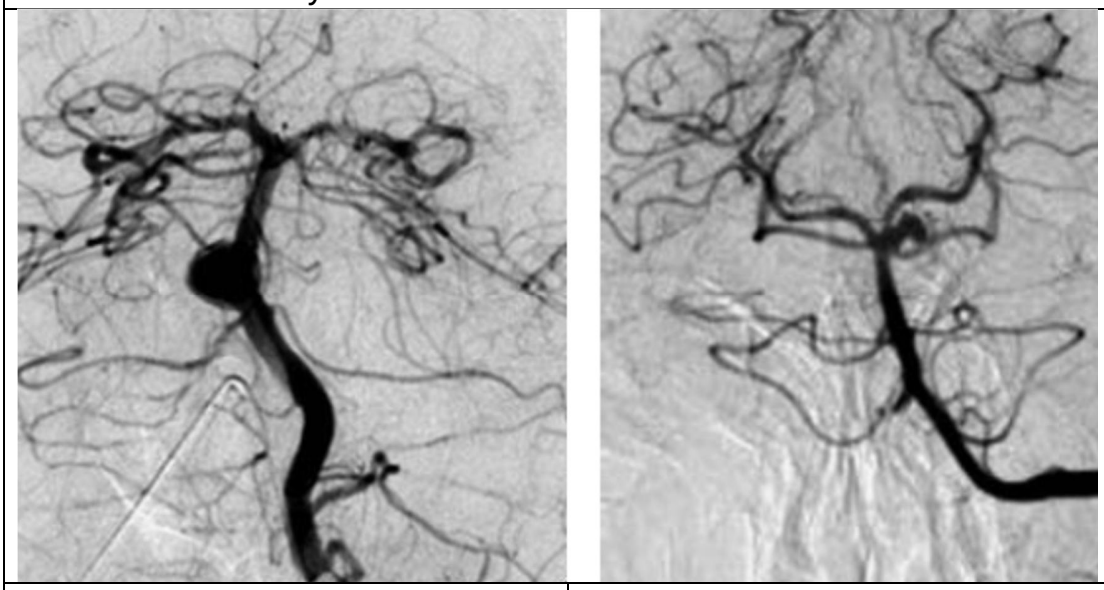


ICA at origin of ophthalmic artery Paraophthalmic aneurysm.

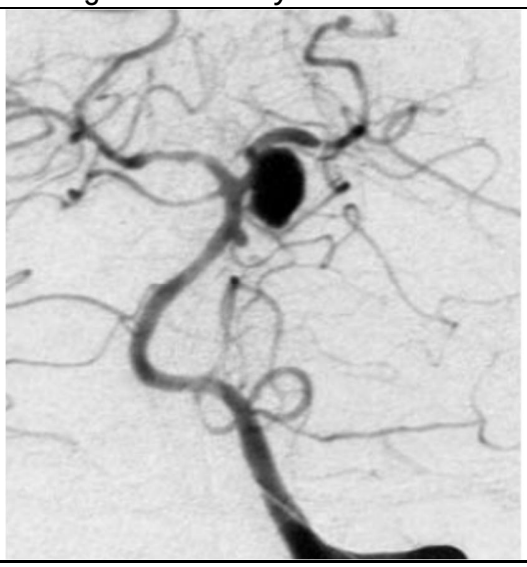


Distal ICA bifurcation aneurysm

Basilar Trunk Aneurysm



P1 segment aneurysm

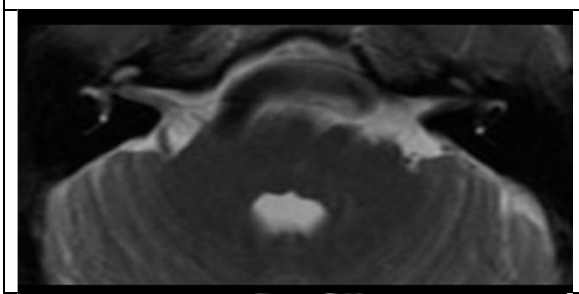


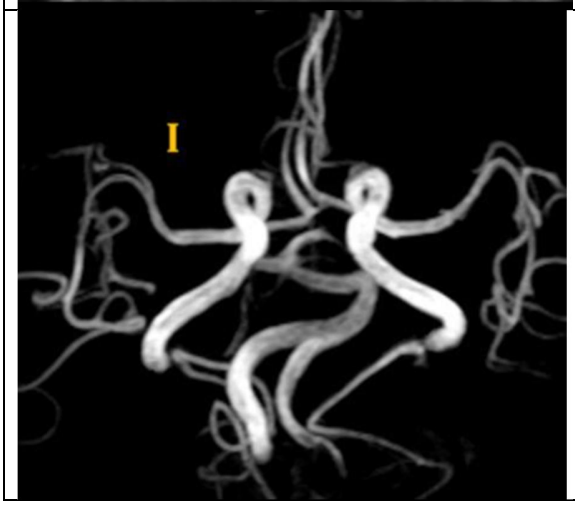
Basilar tip aneurysm

❖ Fusiform Aneurysms

- Segmental arterial dilation without a defined neck.
- Fusiform aneurysms are usually due to atherosclerosis
- Location: Vertebrobasilar system > Anterior circulation
- Imaging finding:
  - Elongated + Dilated + Tortious verteberobasilar artery

• Verteberobasilar Dolichoectasia





❖ Mycotic (Infectious) Aneurysm

- 2%–3% of all intracranial aneurysms.
- A mycotic aneurysm is an aneurysm arising from bacterial infection (bacteremia with septic emboli).

Pathogenesis:

- The vessel wall is digested and a false aneurysm forms which is unstable and highly prone to rupture.

Risk factors:

- Infective endocarditis (Most common cause)
- Intravenous drug use
- Immunosuppression
- Iatrogenic aortic trauma
- Pre existing atherosclerotic plaque or native aneurysm
- Prosthetic arterial devices (stents, grafts)

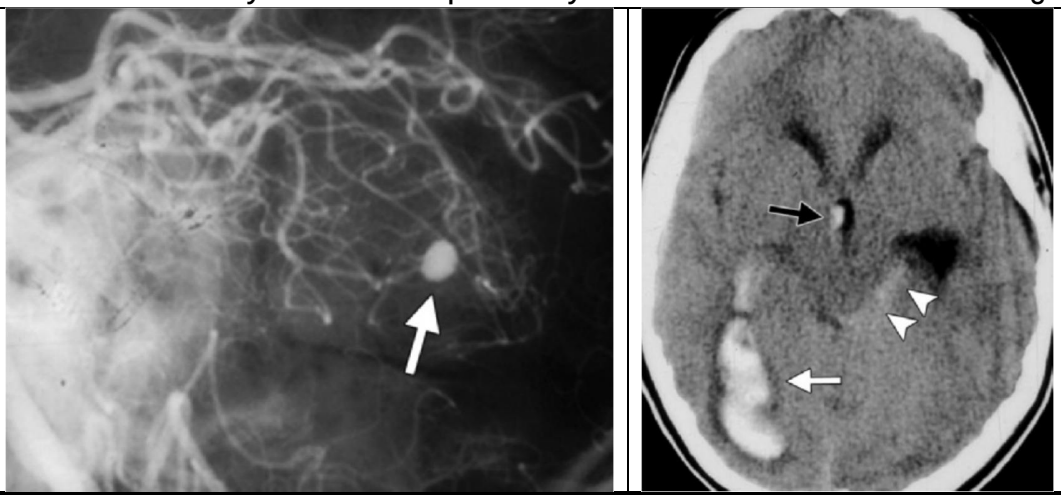
Causative organism:

- Streptococcus - Staphylococcus - Enterococcus

Imaging findings:

- Location: Distal arterial circulation, beyond the circle of Willis.
- Number : Multiple
- Shape: Irregular shape
- Type: False aneurysm.
- Fragile and have a high risk of rupture

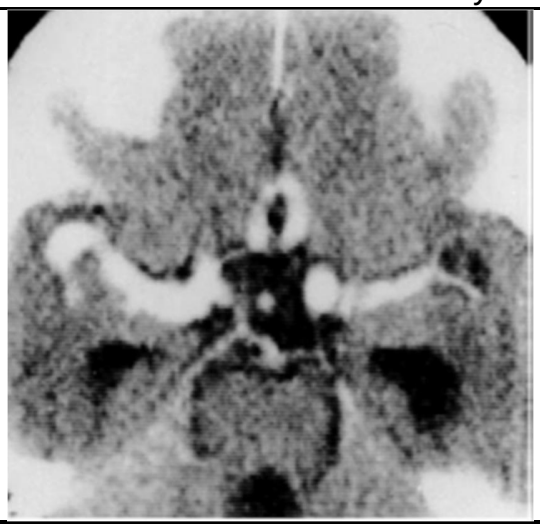
- Rt PCA aneurysm => Intaraparenchymal + SAH + Intraventricular Hge

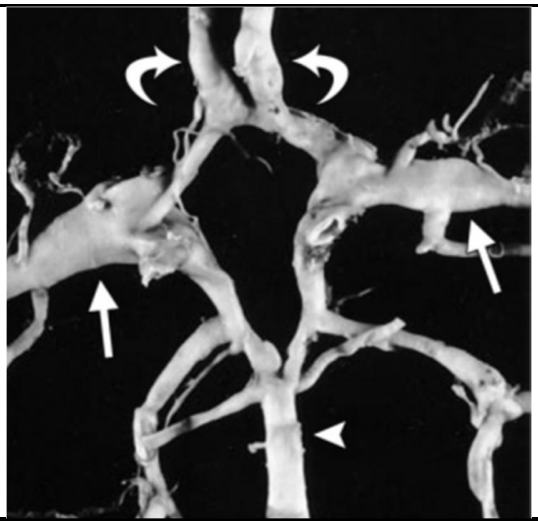


❖Noninfectious Inflammatory Aneurysms

- Adults: known complications of autoimmune vasculitis (primary angiitis of the central nervous system and systemic lupus erythematosus)
- Children: CNS vasculitides including Kawasaki disease
- Acquired cerebral aneurysmal arteriopathy of HIV infection
- Pathogenesis: Noninfectious transmural inflammation of the arterial wall leading to wall failure and rupture

➢ HIV infection: Fusiform aneurysmal dilatations of MCAs and ACAs





❖ Oncotic (Neoplastic) Aneurysm

- An oncotic aneurysm is an aneurysm caused by neoplasm.
- A benign left atrial myxoma may peripherally embolize and cause a distal oncotic aneurysm.

<u>Pathogenesis:</u>

- Cerebral embolization of neoplastic cells with infiltration of the vessel wall and subsequent aneurysm formation. (as mycotic aneurysm)

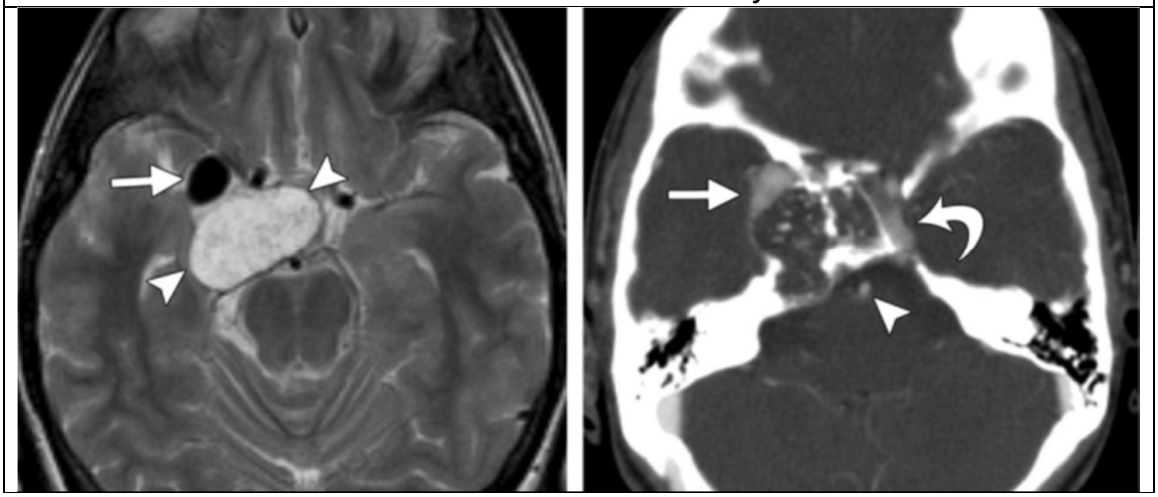
<u>Hematogenous spread for original Tumor:</u>

- Cardiac myxoma
- Choriocarcinoma
- Bronchogenic carcinoma
- Renal cell carcinoma
- Undifferentiated carcinoma

<u>Contiguous spread from adjacent tumors</u>

- Chondrosarcoma of the skull base
- Glial malignancy

- Skull base chondrosarcoma => Rt ICA aneurysm



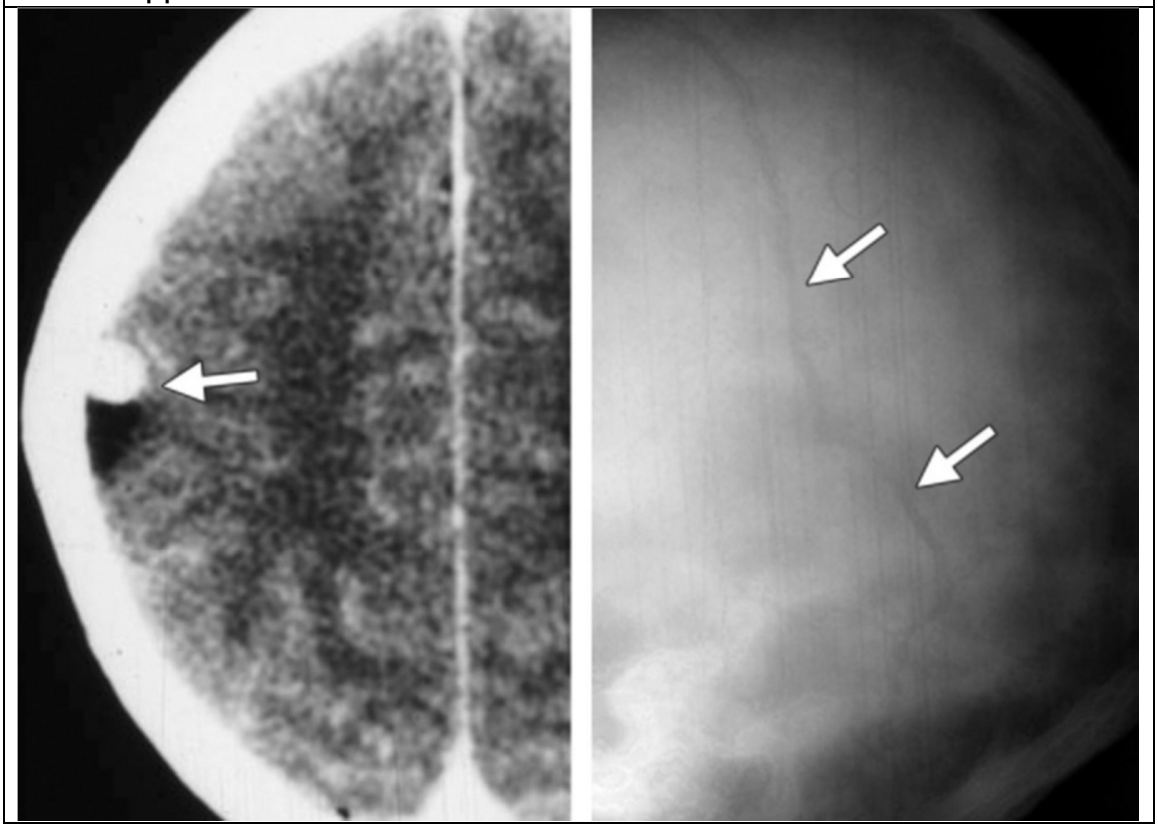
❖ Traumatic Aneurysms

- Pseudoaneurysms = don't contain the typical 3 histologic layers of the vessel wall.

- Trauma is the second most common cause of cerebral aneurysm in the pediatric age group
- < 3% of cerebral aneurysms in adults

<u>Most common sites:</u>

- Pericallosal artery
- Cavernous ICA => Direct carotid–cavernous sinus fistula
- Peripheral aneurysma can occur if the leptomeninges become entrapped in a skull fracture



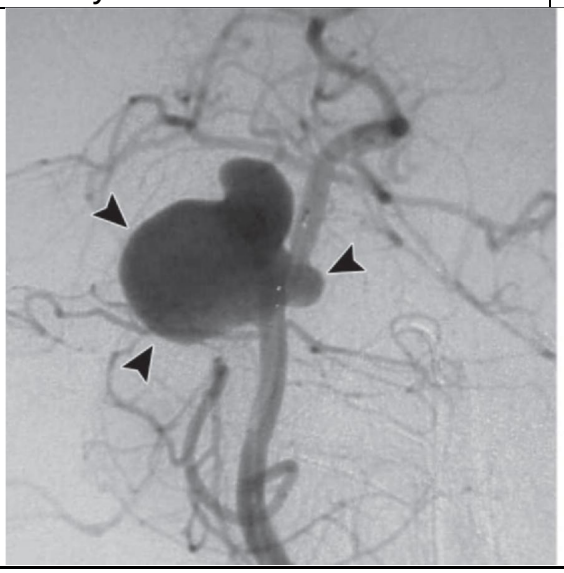
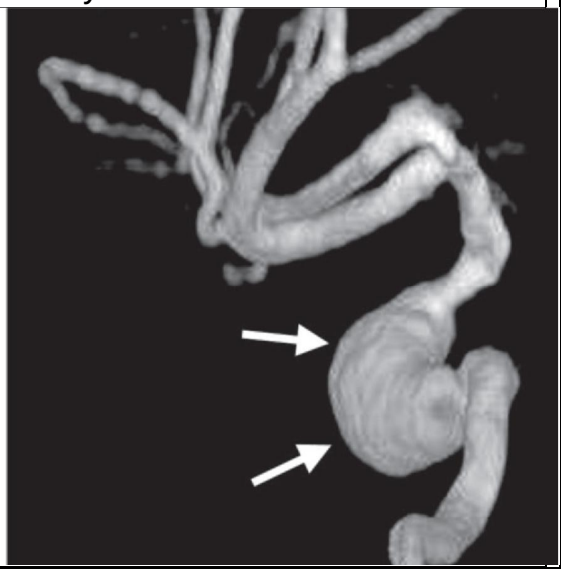
❖ Vasculopathic Aneurysms

- Vasculopathic aneurysms are caused by congenital cerebral aneurysmal arteriopathies
- 10% of cerebral aneurysms in the pediatric population

Congenital Cerebral Aneurysmal Arteriopathies
Collagenopathies and other connective tissue disorders
Osteogenesis imperfecta (type I collagenopathy)
Ehlers-Danlos syndrome type 4 (type III collagenopathy)
Alport syndrome (type IV collagenopathy)
Juvenile Paget disease with osteoprotegerin deficiency
Pseudoxanthoma elasticum
Vascular dysplasias
Tuberous sclerosis
Autosomal dominant polycystic kidney disease
Neurofibromatosis type 1
PHACES* syndrome
Loeys-Dietz syndrome
Marfan disease
Klippel-Trénaunay-Weber syndrome
Alagille syndrome

Other arteriopathies
Growth hormone deficiency
Seckel syndrome (bird-headed dwarfism)
Majewski type II dwarfism
Cutis marmorata telangiectatica congenita
Type IIb hyperlipoproteinemia
Menkes kinky-hair syndrome
Roberts syndrome (pseudo–thalidomide syndrome)

- Large (>12 mm) and often have fusiform morphology
- within or proximal to the circle of Willis

Basilar artery trunk fusiform aneurysm	ICA caverbous segment fusiform aneurysm
	

❖ Aneurysms Associated with AVM

- The incidence of these aneurysms in AVMs is up to 25%
- 50% are located on a feeding artery
- 25% within the nidus
- AVM-associated aneurysms contribute to an increased risk of hemorrhage

❖ Familial Aneurysms

- Saccular IAAs in a sibling pair or in at least three first-degree relatives constitute a clinical entity known as familial intracranial aneurysms (FIAs).

- Epidemiologic studies have shown that 10%– 20% of all individuals with aneurysmal SAH have a family history of intracranial aneurysms,
- FIAs account for only 5% of cerebral aneurysms among patients who present in the first 2decades of life

❖ Idiopathic Aneurysms

- Idiopathic IAAs are a category defined by exclusion.
- IAAs are classified as idiopathic when the affected child has no history of trauma, hemodynamic stressors, known acquired or congenital arteriopathies, infections, or tumors and no family history of IAAs.

- Most cases (~45%) of cerebral aneurysm manifesting in the first 2 decades of life

❖ Complications of Aneurysm:

A) Rupture:

- ⇨ Subarachnoid Hemorrhage:
  - o Hydrocephalus
  - o Vasospasm => Ischemia
- ⇨ Inraparenchymal hemorrhage
- ⇨ Intraventricular hemorrhage

B) Thrombosis:

- ⇨ Emboli => Ischemia

C) Mass effect

- ⇨ Cranial nerve palsy
- ⇨ Headache

D) Rebleeding:

- ⇨ 50% Rebleed within 6m
- ⇨ 50% Mortality

❖ Rupture Risk

- Size most important (but not only) factor
  - o Low rupture risk if < 7 mm
  - o High rupture risk if> 2.5 cm
- Configuration
  - o Multilobed > round/ovoid shape
  - o Apical "tit" or "bleb" present
  - o Aspect ratio (length vs neck) > 1.6
- Perianeurysmal environment (contact with other structures)
- Other (hypertension, female gender, smoking)

❖ Identify Bleeding Aneurysm

1) <u>Location of SAH:</u>

- Anterior interhemispheric fissure SAH
  - ⇨ Acom artery aneurysm (33% of intracranial aneurysms).
- Suprasellar cistern SAH
  - ⇨ Pcom artery aneurysm (33% of intracranial aneurysms).
- Sylvian fissure SAH
  - ⇨ MCA aneurysm (20% of intracranial aneurysms).
- Perimesencephalic cistern SAH
  - ⇨ Basilar tip aneurysm (5% of intracranial aneurysms)
  - ⇨ Benign nonaneurysmal perimesencephalic SAH.
- 4^{th} ventricle hemorrhage
  - ⇨ PICA aneurysm

2) Largest aneurysm is most likely to bleed

3) Irregular lobulated aneurysm is most likely to bleed

4) Contrast extravasation (Rare)

5) Vasospasm adjacent to bleeding aneurysm.

❖ Lines of Treatment

1) Endovascular Coil embolization
2) Surgical Clipping

A) High Flow Vascular Malformations

1) Cerebral Arteriovenous Malformation

- Brain AVMs, congenital high-flow vascular malformation with abnormal connection between cerebral arteries and veins, resulting in arteriovenous shunting with an intervening network of vessels within the brain parenchyma and lack of a true capillary bed

Types according to nidus:

- Compact = Glomerular AVM = No interposed normal brain tissue
- Diffuse = Proliferative AVM = Brain parenchyma is interspersed throughout the tangle of vessels

Types according to location:

- Superficial=Cortical: Cerebral a (ACA-MCA-PCA) + Cortical v
- Deep=Perforators (Lenticulostriate - Thalamostriate) + Deep venous system
- Ventricular= Choroidal a + Deep venous system.

Spetzler-Martin Cerebral AVM grading system (I-V)

- Size of nidus
  - Small (<3cm) = 1
  - Medium (3 - 6cm) = 2
  - Large (> 6cm) = 3
- Eloquence of adjacent brain
  - Non-eloquent = 0
  - Eloquent = 1
- Venous drainage
  - Superficial only = 0
  - Deep = 1

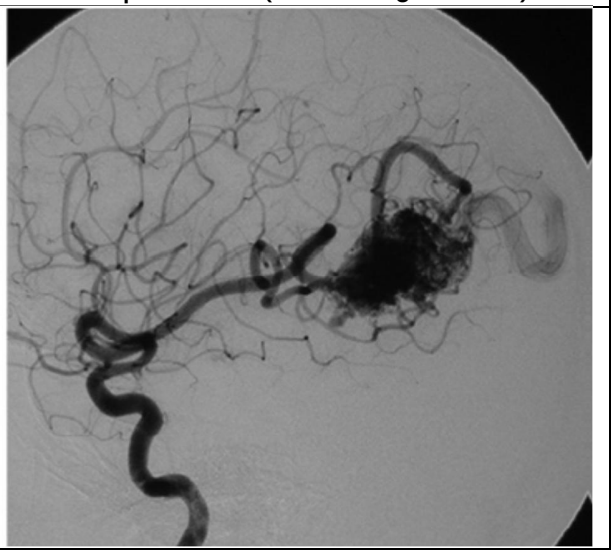
Lines of treatment

- Conservative.
- Open surgery.
- Endovascular treatment: transarterial or transvenous approach: Embolization with liquid embolic agent (Onyx - NBCA) – Coils.
- Stereotactic radiosurgery.
- Combined

Risk of future hemorrhage	Risk of nonhemorrhagic neurologic deficits
• Previous hemorrhage • Intranidal aneurysms • Venous stenosis or ectasia (pouches) • Deep venous drainage • Single venous drainage • Deep or posterior fossa locations	• High-flow shunt • Venous congestion or outflow obstruction • Long pial course of draining vein • Perifocal or perinidal gliosis • Mass effect or hydrocephalus • Arterial steal

Imaging Findings:

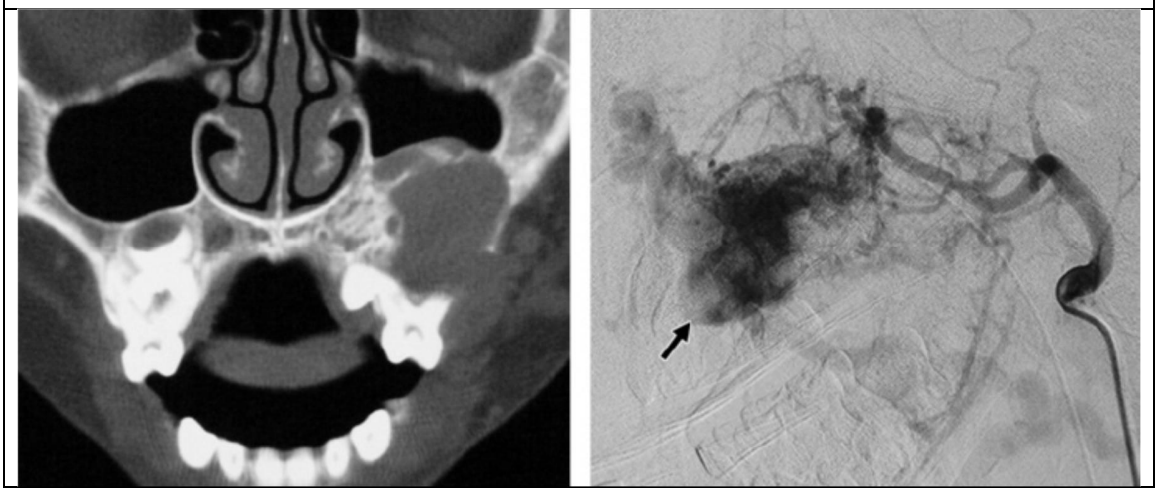
- Vascular nidus ("nest"):
  - Embedded within the brain parenchyma
  - Numerous serpentine vessels:
    - CTA: Strong uniform enhancement
    - MRI: black flow-voids.
- Enlarged feeding artery
- Early venous drainage
- Adjacent brain changes
  - No mass effect unless complicated with hemorrhage
  - Vascular steal and ischemia
  - Gliosis /encephalomalacia (high T2 and FLAIR signal)
  - Dystrophic calcification and blood products (blooming on T2*).



❖ Cerebrofacial Arteriovenous Metameric Syndrome CAMS

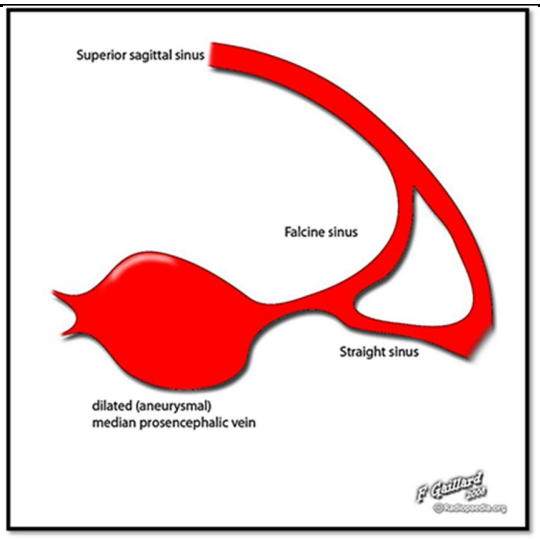
- also known as Wyburn-Mason syndrome or Bonnet-Dechaume-Blanc disease
- Multiple Cerebral + Facial AVM
- CAMS type 1 involves the medial prosencephalon:
  - AVMs at the corpus callosum, hypothalamus (hypophysis), and nose.
- CAMS type 2 involves the lateral prosencephalon:
  - AVMs at the occipital lobe and optic tract, including the thalamus, retina, and maxilla
- CAMS type 3 involves the rhombencephalon:
  - AVMs at the cerebellum, pons, and mandible.

Facial osseous AVM



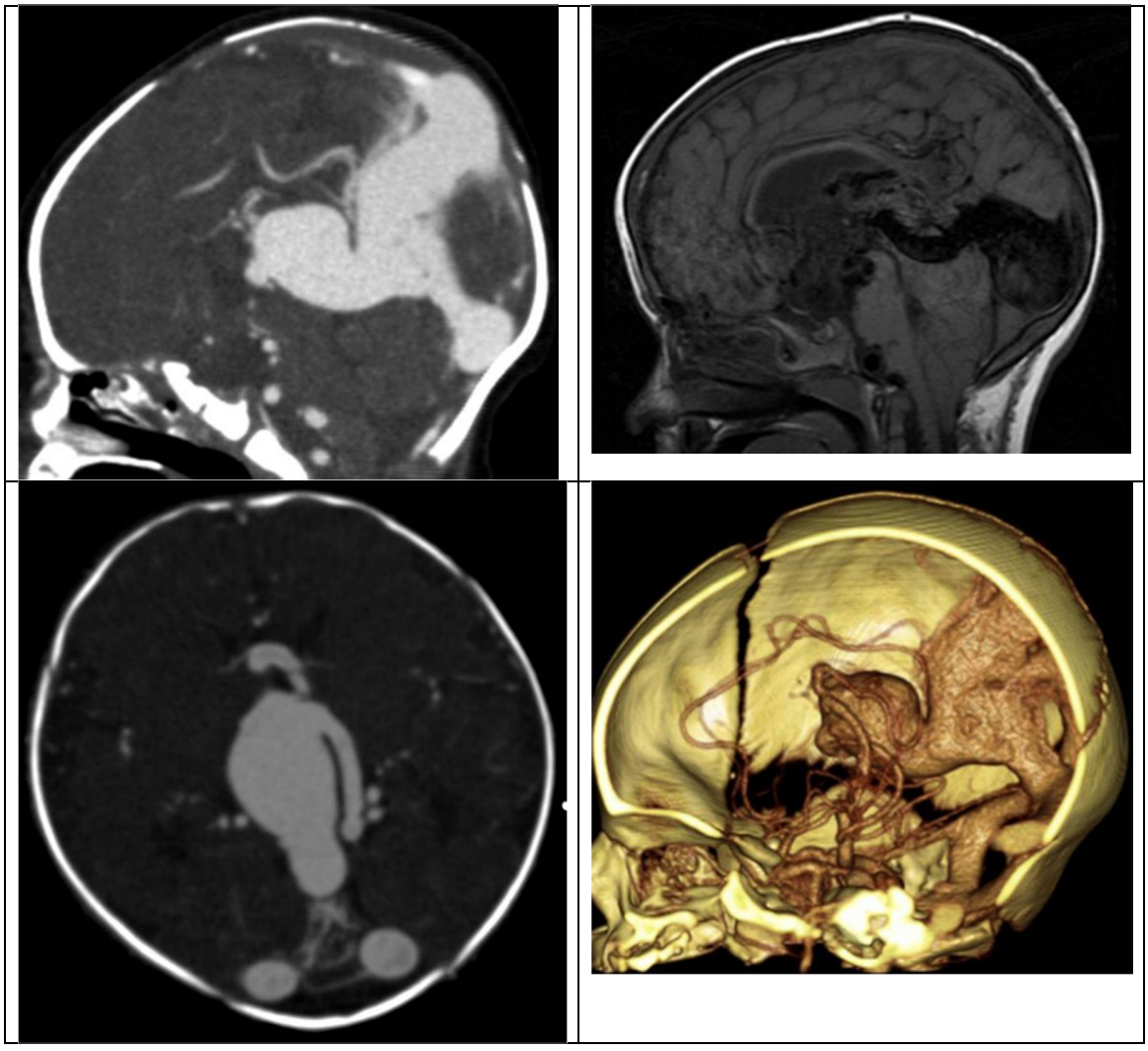
❖ Vein Of Galen Malformation

- Rare congenital anomaly
- 30% of pediatric malformations
- Arteriovenous fistulae from thalamoperforator branches into aneurysmally enlarged median prosencephalic vein
- It is an aneurysmal dilatation and abnormal persistence of the median prosencephalic vein (MPV).



❖ Lasjaunias Classification

Choroidal Type = Neonate (Complex=Secondary=AVM)	Mural Type = Infant (Simple=Direct=AVF)
• Multiple feeders including thalamoperforating, anterior and posterior choroidal and pericallosal arteries • converge on a fistula site at the anterior aspect of the median prosencephalic vein (MPV)	• Fistulae in the wall of the median prosencephalic vein (MPV) • supply may be unilateral • Associated with absence or stenosis of dural sinuses • Associated with stenosis at the level of the jugular foramen
• Present earlier (neonate) with cardiac failure	• Present later (infant) and typically with hydrocephalus



2) Dural Arteriovenous Fistula (dAVF)

- High flow direct arteriovenous shunts between the meningeal arterioles and dural venules.

Dural AVF = Dialted trotous Dural A+V+NO nidus.

❖ Types according to location:

- Carotid-Cavernous fistula
- Transverse-Sigmoid
- Tentorial
- Superior sagittal sinus
- Anterior fossa

❖ Cognard Classification

- Type I: No cortical venous drainage. Lowest risk of bleeding.
- Type IIA: Reflux into dural sinus but not cortical veins.
- Type IIB: Reflux into cortical veins: 10–20% hemorrhage rate.
- Type III: Direct cortical venous drainage: 40% hemorrhage rate.
- Type IV: Direct cortical venous drainage with venous ectasia: 66% hemorrhage rate.
- Type V: Spinal venous drainage. May cause myelopathy.

❖ Borden's classification:

- Borden I= Sinus or meningeal venous drainage + No cortical venous reflux = Benign.
- Borden II= Sinus or meningeal venous drainage + Cortical venous reflux.
- Borden III= Cortical venous reflux only = Malignant

Malignant Dural AVF Complications:

- Venous congestion / hemorrhage => Neurological symptoms
- Intracranial hemorrhage
- Seizeures - Dementia - Altered conscious

Lines of treatment

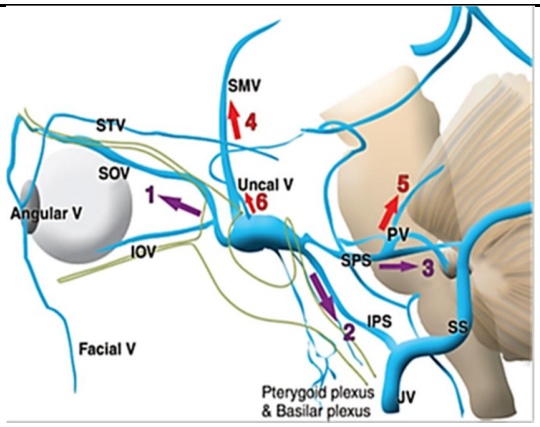
- Conservative.
- Open surgery.
- Endovascular treatment: transarterial or transvenous approach: Embolization with liquid embolic agent (Onyx - NBCA) – Coils.
- Stereotactic radiosurgery.
- Combined

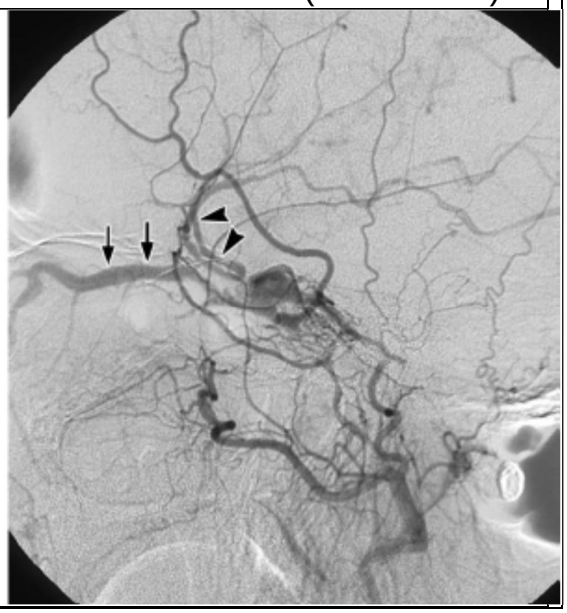
❖ Carotid Cavernous Fistula

Type A: Direct ICA-cavernous sinus high-flow shunt (not true dAVF)
Type B: ICA dural branch - shunt to cavernous sinus
Type C: ECA dural branch - shunt to cavernous sinus
Type D: ECA/ICA dural branches - shunt to cavernous sinus

Venous drainage of cavernous sinus dural AVFs.

1) Anterior drainage:
→ Superior ophthalmic vein (SOV)
→ Inferior ophthalmic vein (IOV)
→ lead to ocular symptoms (eg, exophthalmos and chemosis)
2) Posteroinferior drainage:
→ Inferior petrous sinus (IPS), basilar plexus,
→ Pterygoid plexus
→ lead to bruit and cranial nerve deficits
3) Posterior drainage
→ Superior petrous sinus (SPS)
→ lead to bruit
4) Superior drainage:
→ Cortical reflux into sphenoparietal sinus and superficial middle cerebral vein (SMV)
→ lead to venous infarction and hemorrhage
5) cerebellar (spinal) drainage:
→ into petrous vein (PV),
→ lead to ataxia and hemorrhage
6) Deep drainage
→ into deep middle cerebral
→ Lead to hemorrhage.



Cavernous sinus dural AVF shows multiple flow voids in the left cavernous sinus (arrows).	Normal cavernous sinus
Cavernous sinus dural AVF drain into the inferior petrosal sinus (arrows) and pterygopharyngeal plexus (arrowheads).	Cavernous sinus dural AVF drains into the superior ophthalmic vein (arrows) and the superficial middle cerebral vein (arrowheads).
	

B) Low Flow Vascular Lesions

1) Cavernous Malformation = Cavernoma = Cavernous Angioma

- Benign sporadic low pressure vascular malformation

Location:

- 60 – 80% Multiple => Inherited disorder called familial cavernomatosis
- 60 – 80% Supratentorial

Clinical picture:

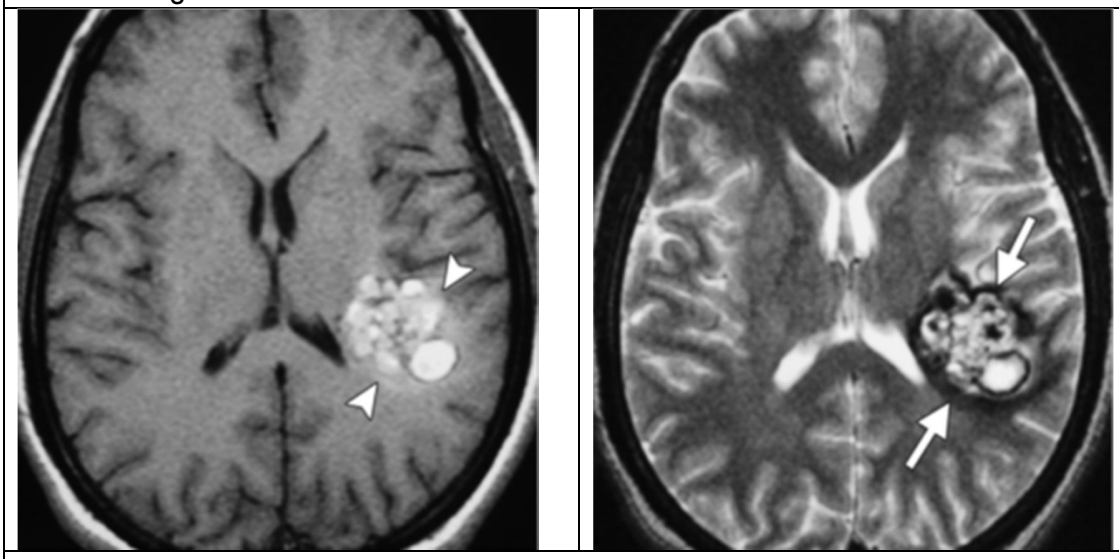
- Seizures
- Neurological deficit
- Headache

- Cavernous malformation is often associated with an adjacent developmental venous anomaly (DVA).
- There is increased risk of bleeding if a DVA is present.
- However, the DVA itself does not have any bleeding risk.

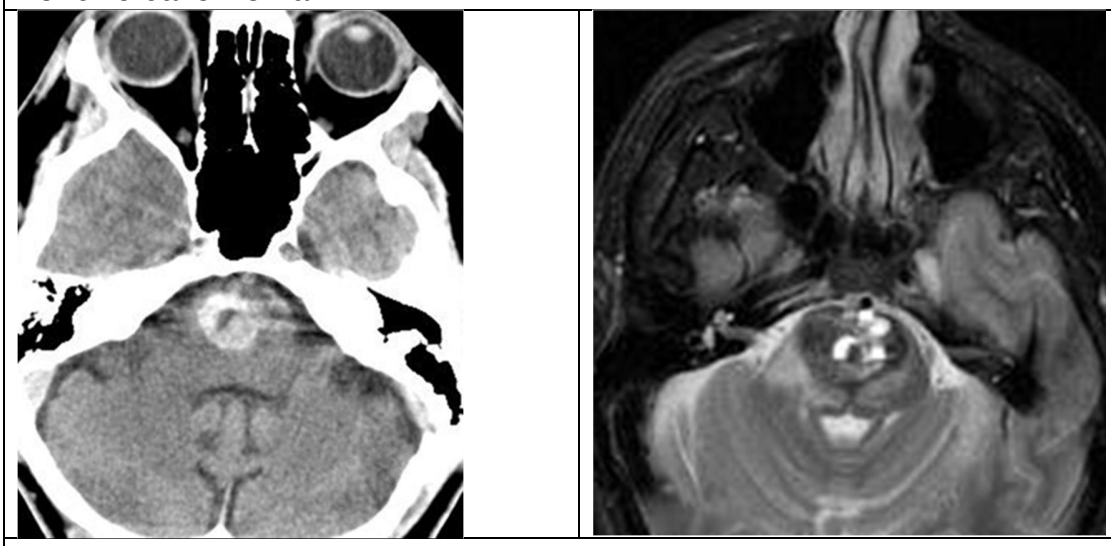
Imaging findings:

- Angiography / CTA: Normal
- CT: Calcifications (Common)
- MRI: Popcorn like lesion:
  - T1&T2: Mixed high and low signal core
  - Low signal rim of hemosiderin
  - T2*: Bloom artifact to detect multiple lesions
  - +C: No enhancement
  - No surrounding brain edema unless complicated

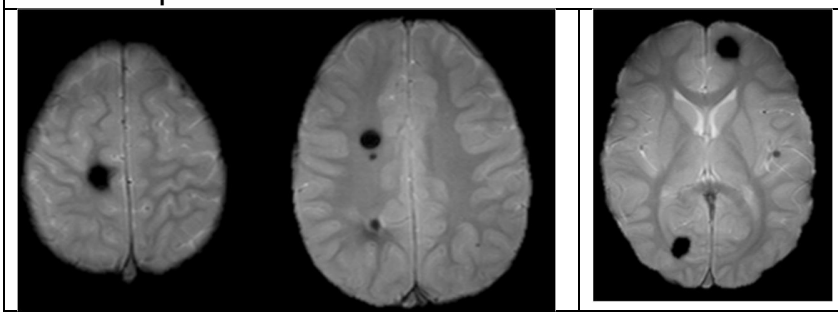
Hemorrhagic cavernoma



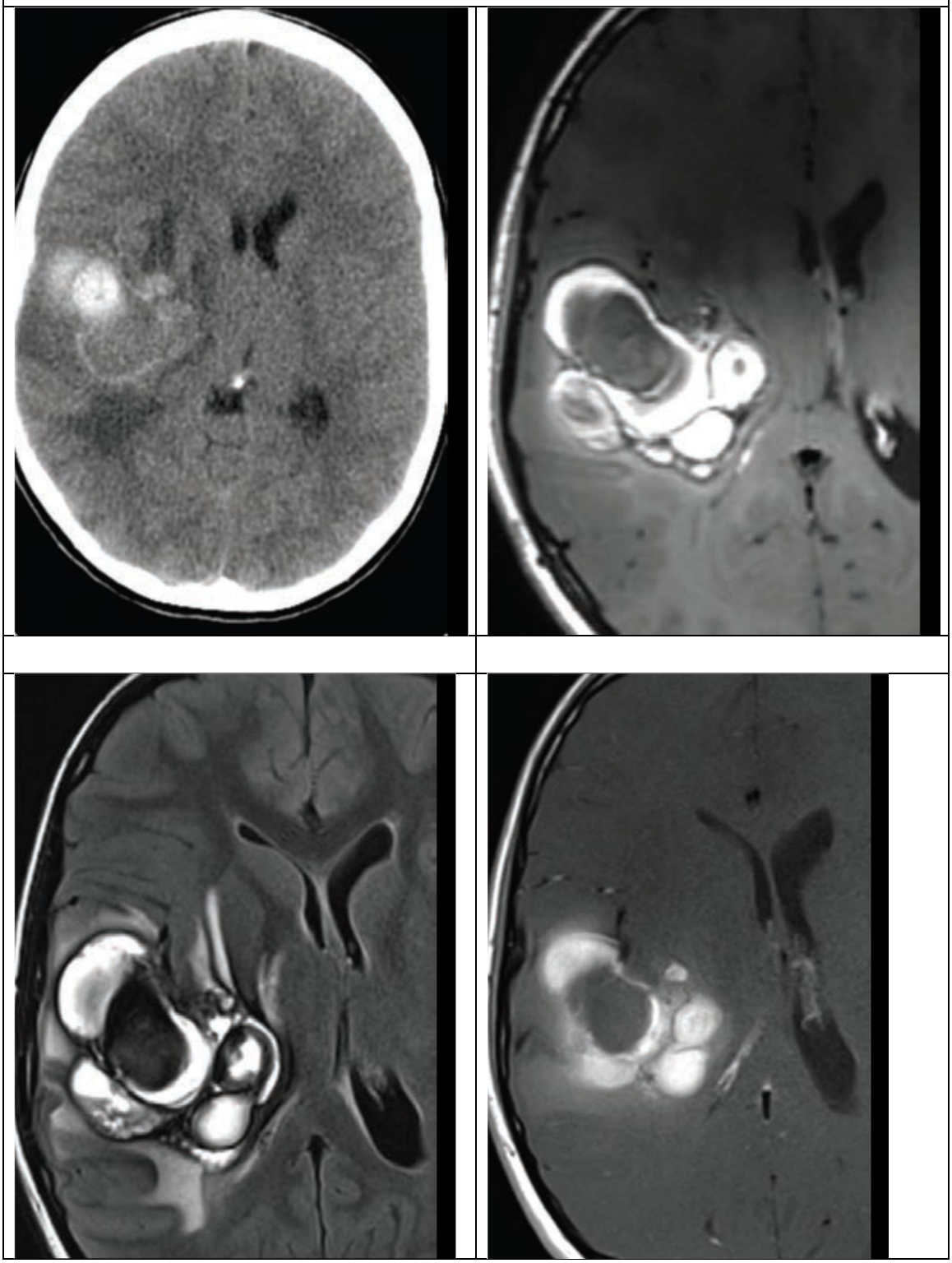
Pontine Cavernoma



T2* Multiple cavernomas



Giant Cavernous Malformation



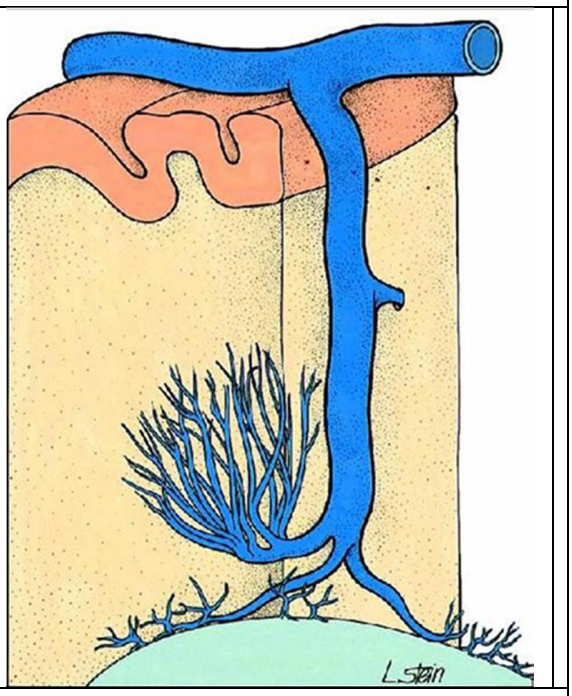
2) Developmental Venous Anomaly (DVA) = Venous Angioma

- An abnormal vein that provides functional venous drainage to normal brain

Location:

- Supratentorial (65%): Frontal is most common site; adjacent to frontal horn of lateral ventricle
- Infratentorial (35%)

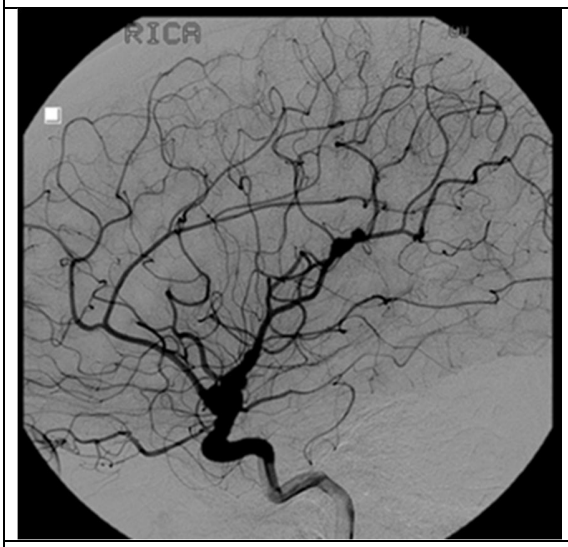
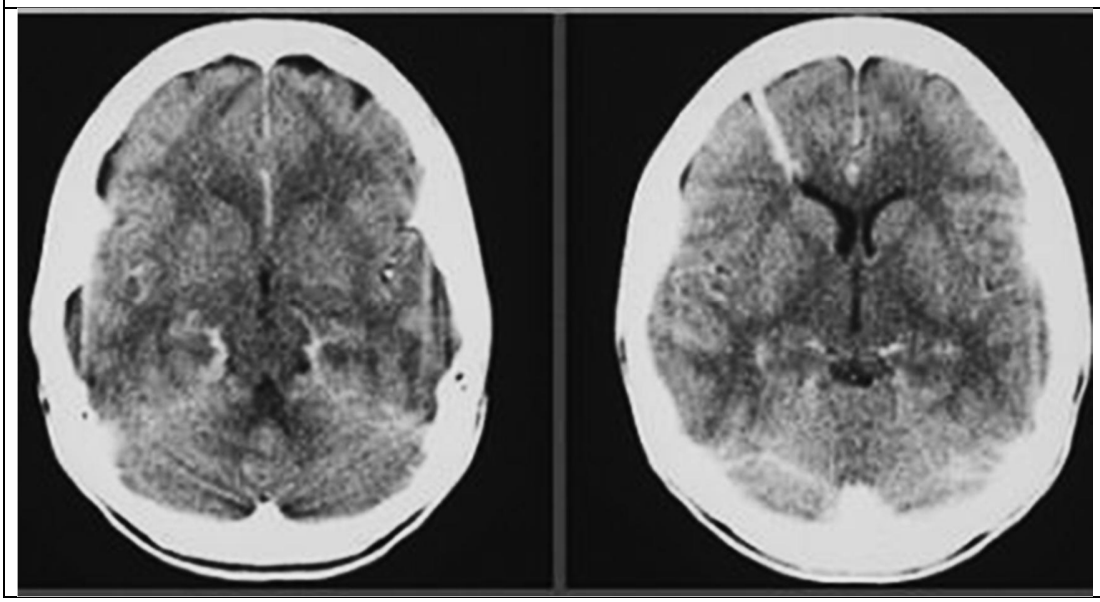
- Umbrella like collection = Caput medusa of medullary veins at the angle of ventricle join to form large collector vein
- The collector vein drains into dural sinus
- 15-20% associated with cavernous malformation
- Most commonly asymptomatic

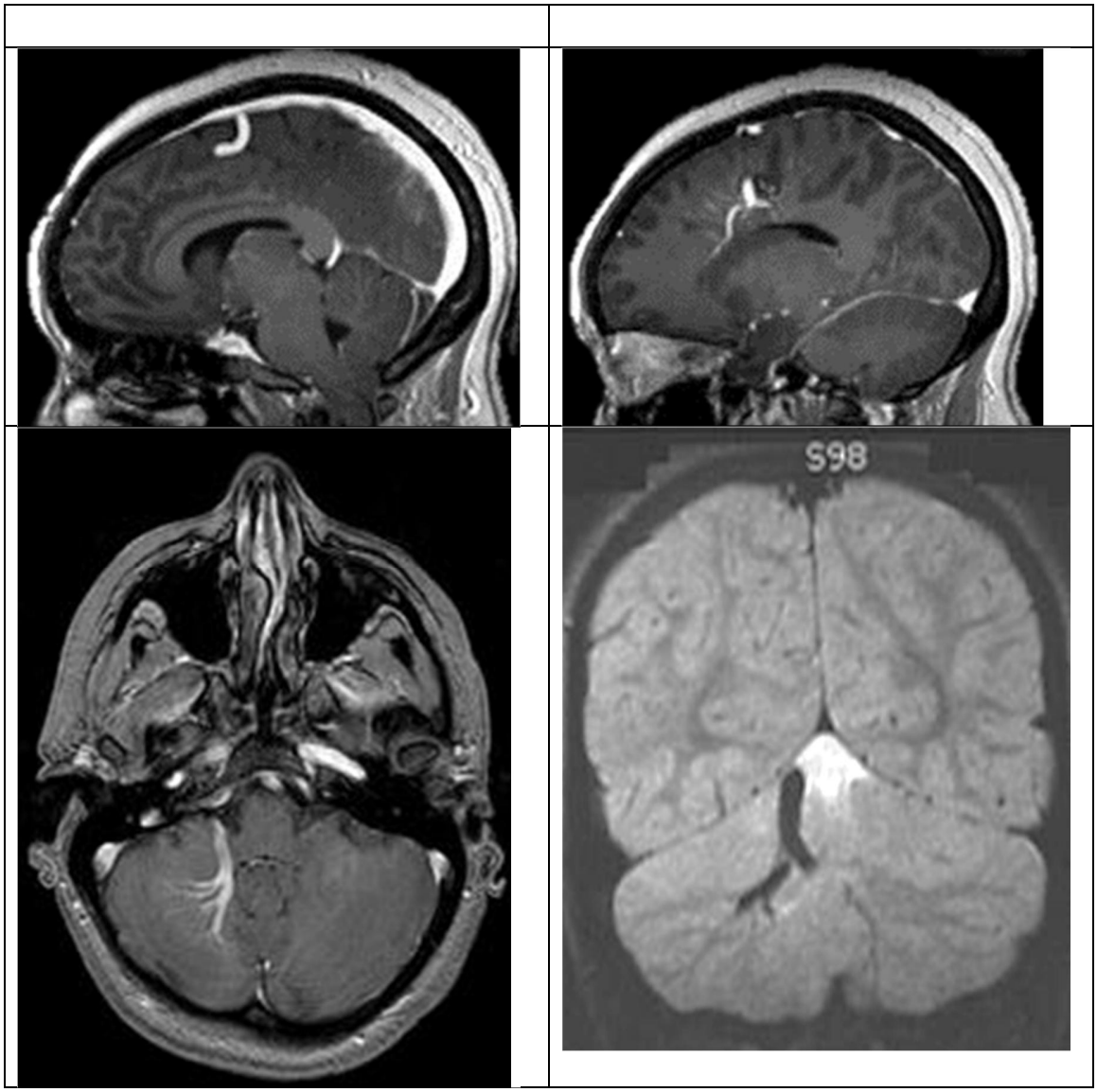


Imaging Findings:

- Angiography / CTA:
  - Arterial phase: Normal
  - Venous phase:
    - Dilated medullary vein = Caput medusa
    - Enlarged transcortical draining vein
- MRI: Linear signal voids
- Normal intervening brain tissue

❖ DVA is a Do Not Touch lesion.

Arterial phase of angiography is normal	Venous phase of angiography shows multiple medullary veins draining into dilated frontal cortical vein draining into superior sagittal sinus
	
CECT: Linear enhancing structure in the frontal lobe coursing between the frontal horn of lateral ventricle and inner table of skull.	
	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3) Capillary Telangiectasia

- is an asymptomatic vascular lesion composed of dilated capillaries with interspersed normal brain.

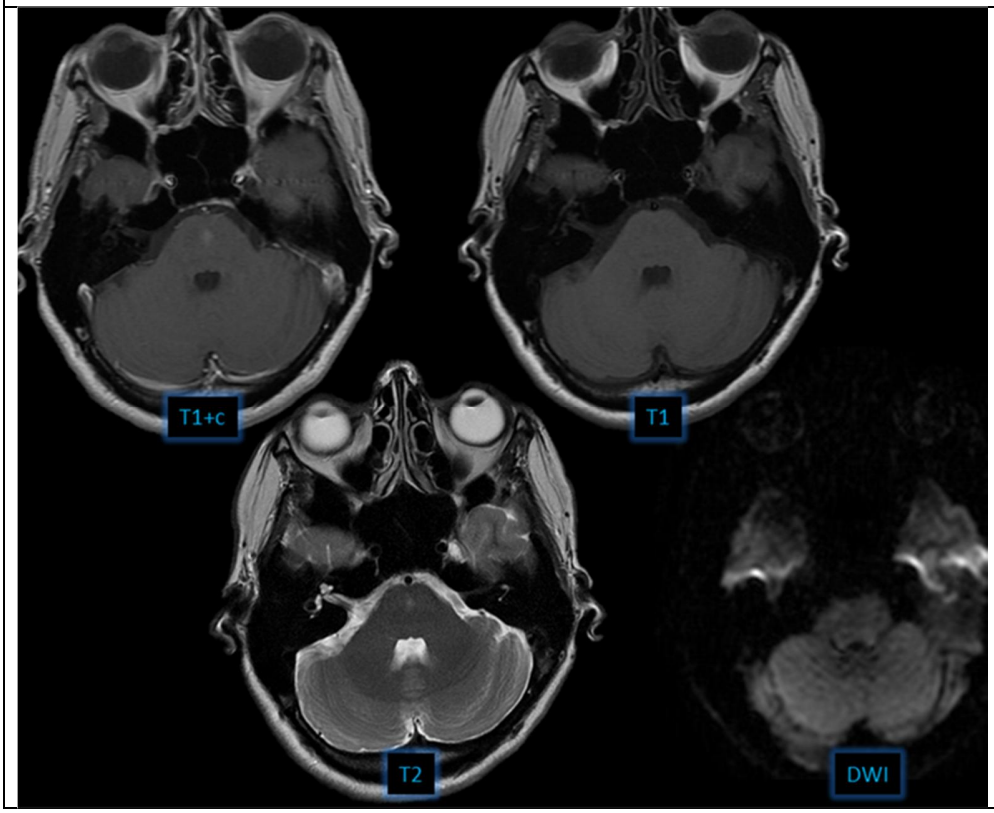
Location:

- Pons – Medulla – Spinal cord

Imaging findings:

- Angiography / CTA: Normal
- T2: Hyperintesne focus
- +C: Faint, brush-stroke-like enhancing focus in the brainstem or pons
- No mass effect or surrounding edema.
- T2* / GRE: may show blooming due to susceptibility.

❖ Capillary telangiectasia is Do Not Touch lesion.

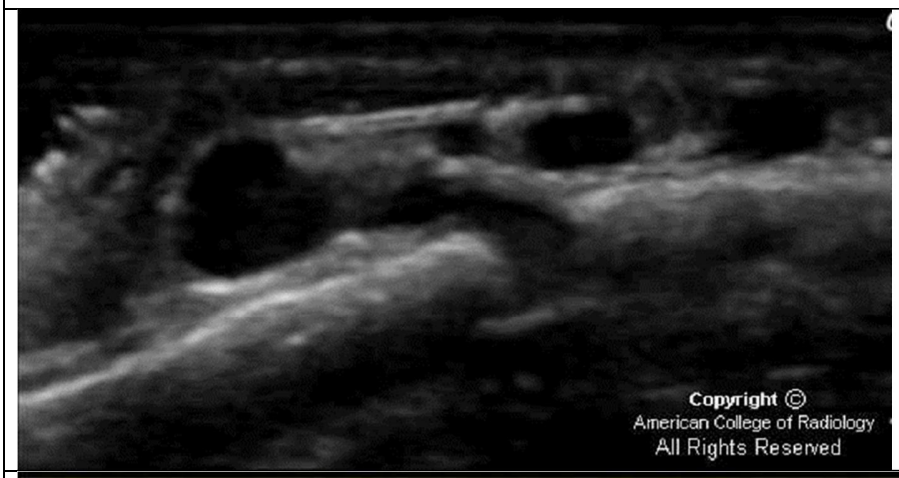


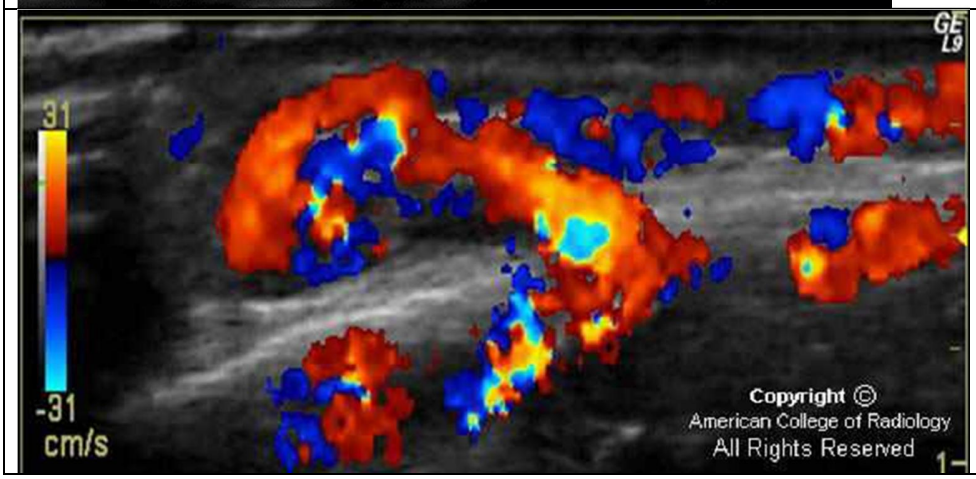
4) Sinus Pericranii

- is an unusual venous anomaly characterized by dilated extracranial veins that communicate with the dural sinuses via transcalvarial emissary veins.

- Doppler sonography is the ideal imaging modality, however, other modalities can be used to diagnose sinus pericranii

- Clinically, patients present with a fluctuant mass that characteristically enlarges with increased intracranial pressure such as crying, a Valsalva maneuver, or dependent positioning.
- Patients tend to be asymptomatic





PERSISTENT CAROTID-VERTEBROBASILAR ANASTOMOSES

- A number of anterior–posterior carotid to basilar connections are formed during embryogenesis.
- Fetal circulation connections normally regress before birth.
- Occasionally, a fetal carotid–basilar connection may persist after birth.

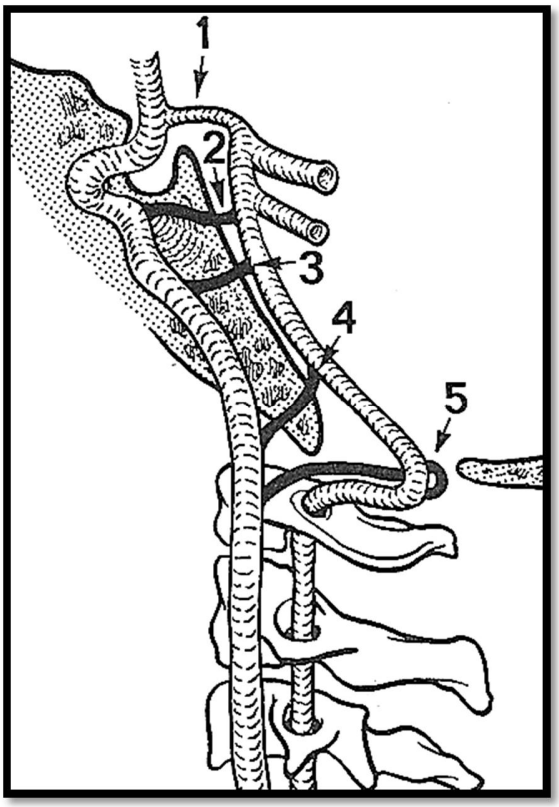
1) <u>"Fetal" origin of PCA:</u>
PCom: ICA → PCA
PCA arises from PCom

<u>2) Primitive Trigeminal Artery:</u>
Cavernous ICA → Basilar
courses adjacent to the trigeminal nerve
1st most common (0.1-0.5%)

<u>3) Persistent Otic Artery:</u>
Petrous ICA → Basilar
Through IAC
Very rare

<u>4) Persistent Hypoglossal Artery:</u>
Cervical ICA (C1,2) → Basilar
through hypoglossal canal
2nd most common (0.1%)

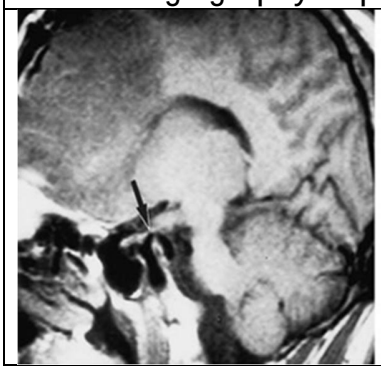
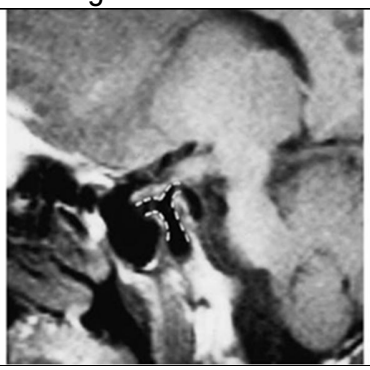
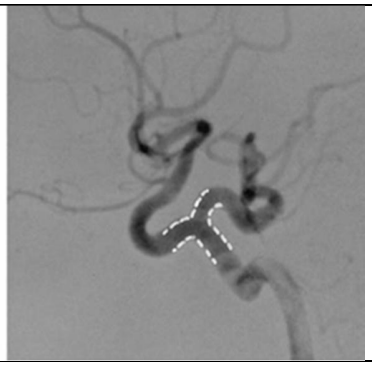
<u>5) Proatlantal Intersegmental Artery:</u>
Cervical ICA (Suboccipital) → Vertebral



Persistent Trigeminal Artery (PTA)

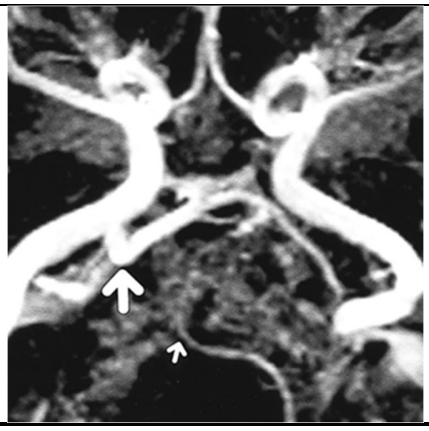
❖ <u>Characteristic trident or tau sign:</u>

Lateral angiography or para-sagittal T1:

❖ Saltzman Classification:

Saltzman type I	Saltzman type II
• PTA supplies distal vertebrobasilar system • PCom usually absent • BA below anastomosis usually hypoplastic	• PTA fills superior cerebellar arteries • Posterior cerebral arteries supplied via patent PCom

Lateral-type PTA	Medial-type PTA
	• Intra-sellar PTA • Trans-hypophyseal PTA
• Courses posterolaterally • Along the trigeminal nerve	• Courses posteromedially. • Compress the pituitary gland • Penetrates the dorsum sellae
	

Cerebral Infection

I. Bacterial

II. Granulomatous

III. Fungal

IV. Parasitic

V. Viral

VI. AIDS

VII. TORCH

VIII. CJD

I- Pyogenic Infection

A- Acute Bacterial Meningitis

Causative Organisms

- Neonates: group B streptococcus
- Children: H. influenza
- Adults: Streptococcus pneumoniae

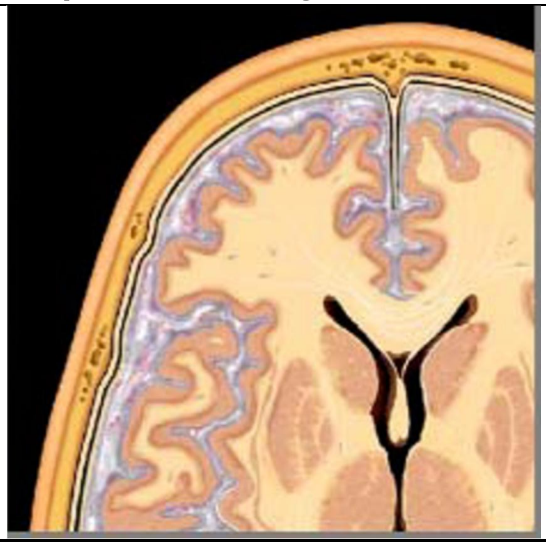
Pathogenesis

- Hematogenous seeding: choroid, leptomeninges
- Contiguous spread from sinusitis, mastoiditis
- Neonatal meningitis - maternal GU infxn, PROM

Clinical:

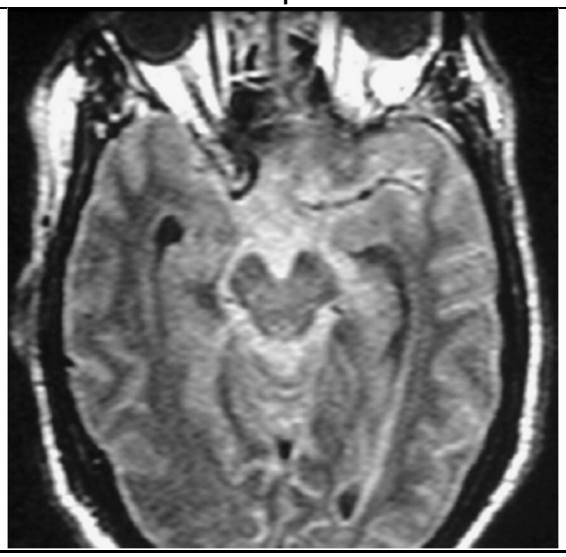
- Neck stiffness, photophobia, cranial nerve dysfunction, lethargy

❖ 2 Patterns of Meningitis

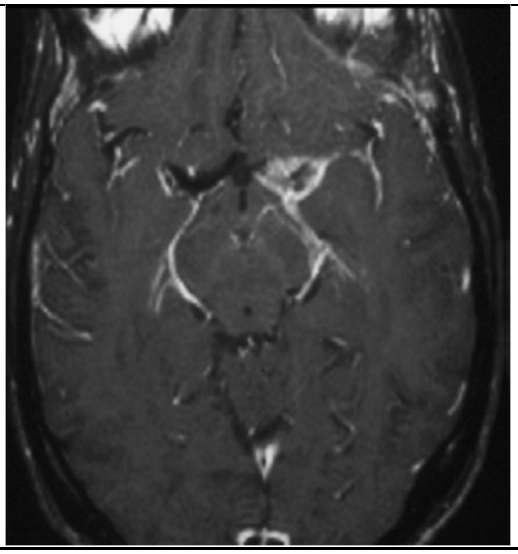
Pachymeningitis	Leptomeningitis
	

❖ Imaging Findings:

- FLAIR: Increased CSF signal in subarachnoid space



- Post Gd: Leptomeningeal enhancement



Complication:

- Hydrocephalus (especially communicating)
- Subdural effusions, empyema
- Infarction: Arterial / Venous
- Vasculitis
- Cerebritis or abscess
- Ventriculitis /ependymitis

B- Bacterial Cerebritis

Etiology

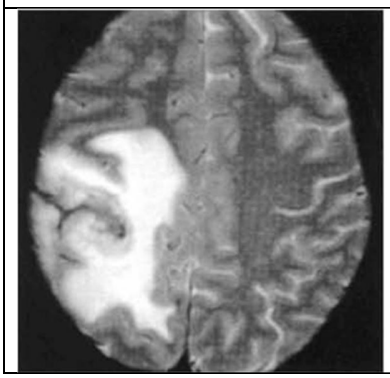
- Direct spread (sinus, mastoid, odontogenic)
- Hematogenous
- Surgery or trauma
- 25% - no source found

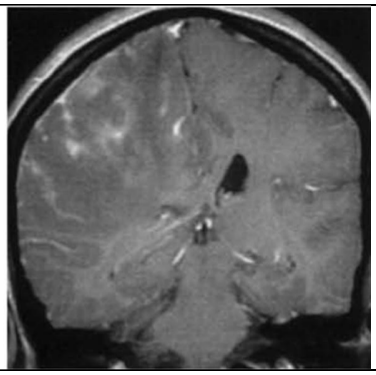
❖ 4 Stages

i) Early Cerebritis	iii) Early capsule
• 3-5 days after infection • T2/FLAIR: high SI +/- minimal enhancement	• 2-4 weeks • Collagenous capsule +/- daughter abscesses • Mass effect
ii) Late cerebritis	**iv) Late capsule**
• 4-14 days after infection • T2/FLAIR: high SI and enhancement	• Weeks/months • Rarely seen as patients are treated earlier • Thick capsule

❖ Imaging Findings:

- T1: patchy area of hypo-intense signal
- T2: patch area of hyper-intense signal
- T1+C: small areas of patchy enhancement.
- Perifocal edema
- Mild mass effect





C- Pyogenic Abscess

Rout of infection:

- Hematogenous dissemination
- Direct spread from paranasal sinusitis or mastoiditis
- Complication of bacterial meningitis

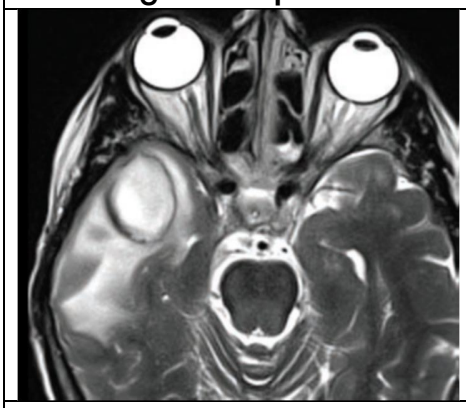
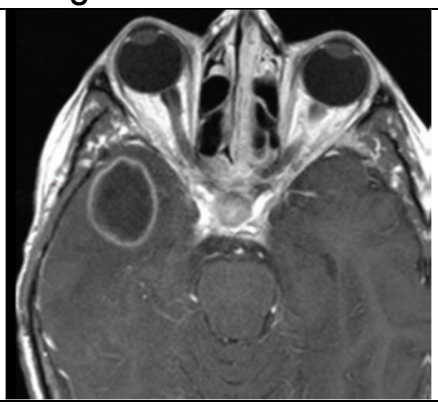
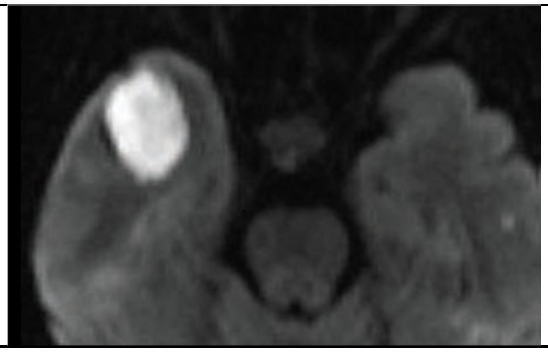
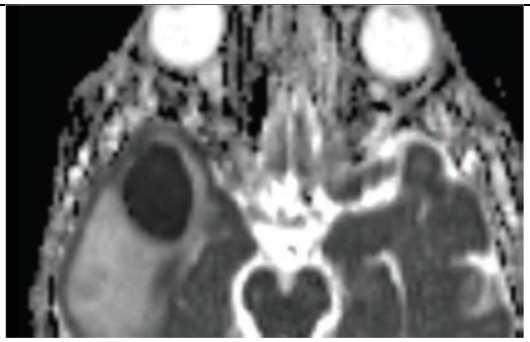
Causative organism:

- Streptococcus, Staphylococcus
- Infants – Citrobacter, Proteus, Pseudomonas, Serratia

- An abscess evolves over four stages (early cerebritis → late cerebritis → early abscess →late abscess)
- Takes about two weeks to fully develop

3 Zones:

- The abscess cavity: low TI and high T2 signal
- The capsule: low signal in both T1 and T2.
- Peripherally, a zone of edema is usually present surrounding the lesion.

Low signal capsule	Ring enhancement
	
Restricted diffusion	**Low ADC**
	

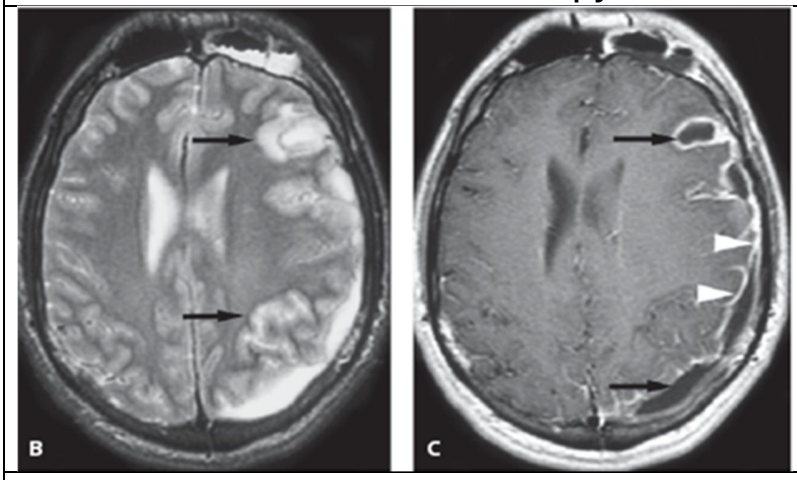
D- Empyema

- is a loculated collection of pus within a membrane-bound space.
- can occur either in the subdural or epidural space or may involve both spaces simultaneously.
- Subdural >> Epidural empyema.

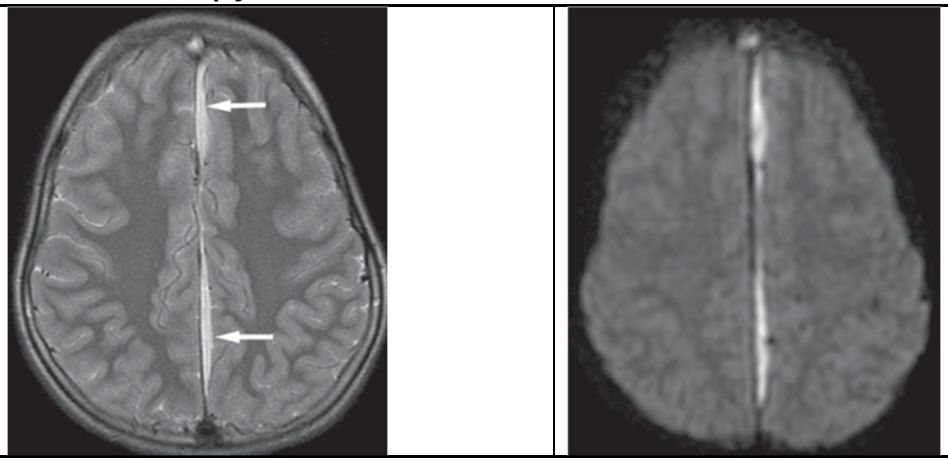
Source of Infection:

- Paranasal sinus infection.
- Mastoiditis
- Osteomyelitis
- Intracranial surgery.
- In infants, meningitis can lead to subdural collection.

Frontal Sinusitis => Subdural Empyema



Subdural Empyema => Restricted Diffusion



E- Ventriculitis

Imaging findings:

- Cerebral abscess
- Ventricular dilatation
- T1: dependent hypointense debris (inflammatory exudate / pus)
- T1+C: Ventricular wall enhancement
- DWI: Restricted diffusion of the dependent debris / pus

Axial T1+C	Axial DWI

II- CNS TB

Causative Organism:

- M. tuberculosis : mostly
- M. bovis rare

Risk factors:

- HIV
- IV drug abuse
- Immune compromised states,
- Homelessness,
- Crowded conditions in confined populations (e.g., prisons and nursing homes),
- Immigration from endemic areas.

Mode of transmission:

- Hematogenous spread from lung (common)
- Direct spread from mastoid nearby infection (rare)

❖ Patterns of infection:

1) Meningitis.
    a. Leptomeningitis / pachmeningitis
    b. Hydrocephalus
    c. Vascular Complication
    d. Cranial Neuropathy
2) Tuberculoma / Granuloma.
3) Miliary TB
4) Cerebral Abscess.
5) Focal Cerebritis.
6) TB Encephalopathy
7) Hypophyseal tuberculoma
8) Calvarial Osteomylitis + Epidural Abscess
9) Spinal cord:
    a. Arachnoiditis
    b. Focal intramedullary tubercloma

1) TB Meningitis

- TB meningitis is the most common cause of chronic meningitis in developing countries.
- Tuberculous meningitis may manifest in two forms:
    - Leptomeningitis: most common.
    - Pachymeningitis: less common

i) Leptomeningitis (more common):

- Basal cisterns (Interpeduncular + Prepontine + Suprasellar + Perimesencephalic)
- + extend along sulci covering sylvian fissures and convexities
- Follow cortical gyri and sulci

ii) Pachymeningitis (less common):

- Follow inner surface of calvarium
- Extend along falx and tentorium

iii) Hydrocephalus

- Basal TB meningitis => obstruction of the subarachnoid space at the basal cisterns by inflammatory exudate => Communicating obstructive hydrocephalus.

iv) Vascular Complication

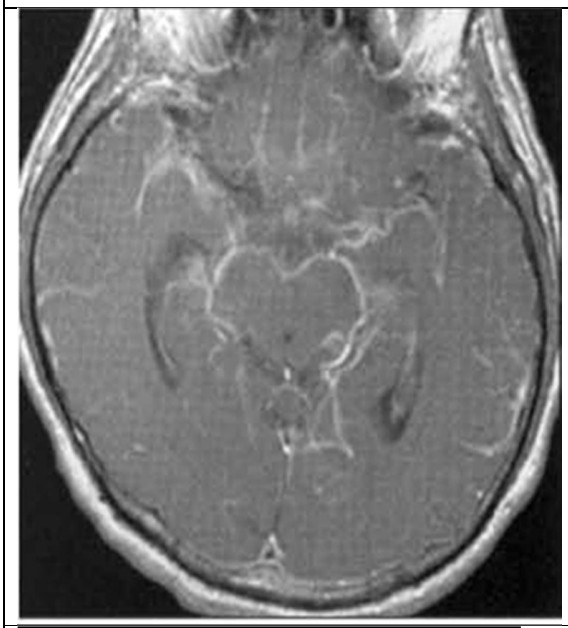
- Basal TB meningitis => Subarachnoid space inflammatory exudate => TB arteritis / vasculitis of medium and small sized arteries => Vascular occlusion => Cerebral Infarction + Infarction of BG and IC.

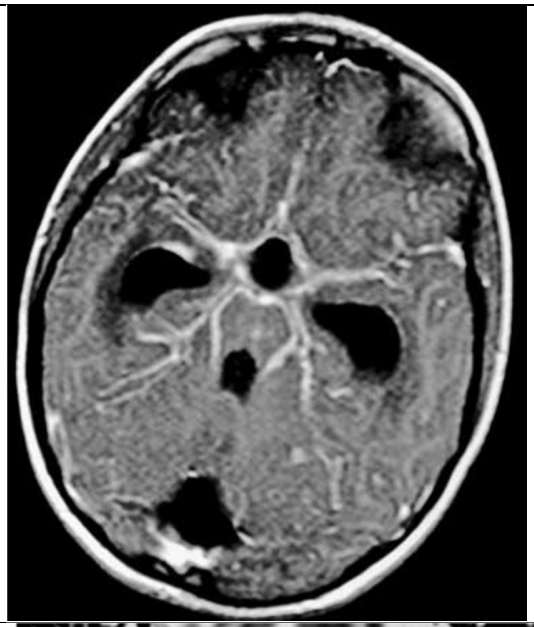
v) Cranial Neuropathies

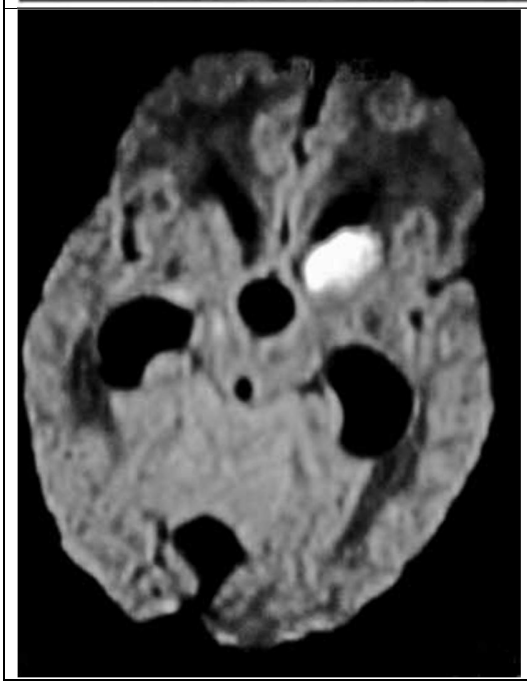
Cranial neuropathies are thought to be due to

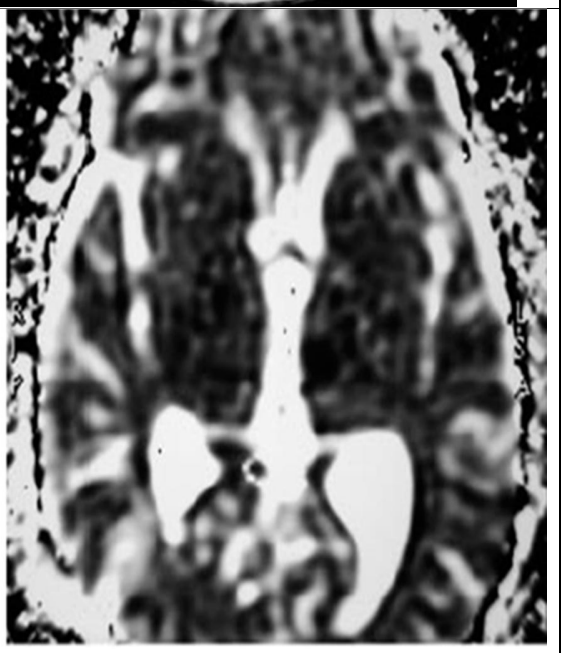
- Vascular compromise resulting in ischemia of the nerve
- Entrapment of the nerves by the exudate.
- Nuclear and supranuclear involvement of the cranial nerve nuclei in the brainstem due to infarct, focal cerebritis, or tuberculoma

Triad = Basal Leptomeningitis + Hydrocephalus + BG Infarction









2) CNS Tuberculoma

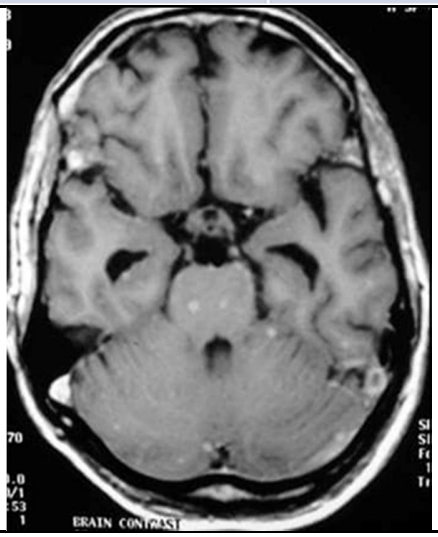
- Single or multiple
- With or without meningitis.
- Anywhere
- Neurological deficit according to the topographic location.

- MRI signal vary according to the stage of maturation:
  - Noncaseating
  - Caseating with solid content
  - Caseating with liquid content

	NON CASEATING GRANULOMA	SOLID CASEATION	LIQUID CASEATION
T1W	Hypo	Hypo to iso with slightly hyperintense rim	HYPO
T2W	**HYPER**	**HYPO**	Central **HYPER** with **HYPO**.. rim
Enhancement	**NODULAR** Homogenous	**RIM**	**RIM**

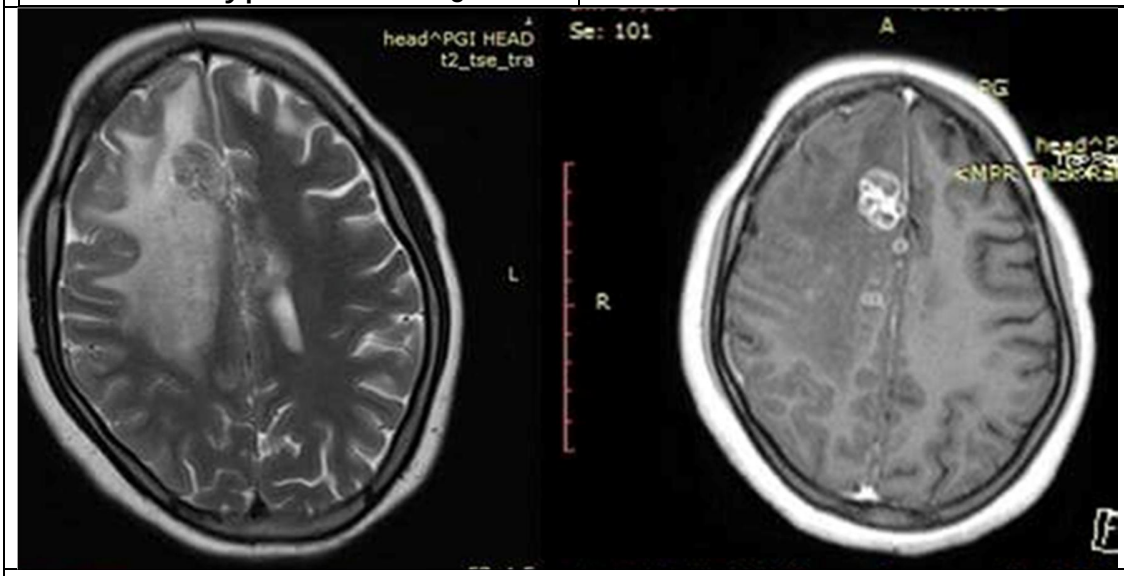
<u>Non-caseating tuberculoma</u>

Nodular enhancement



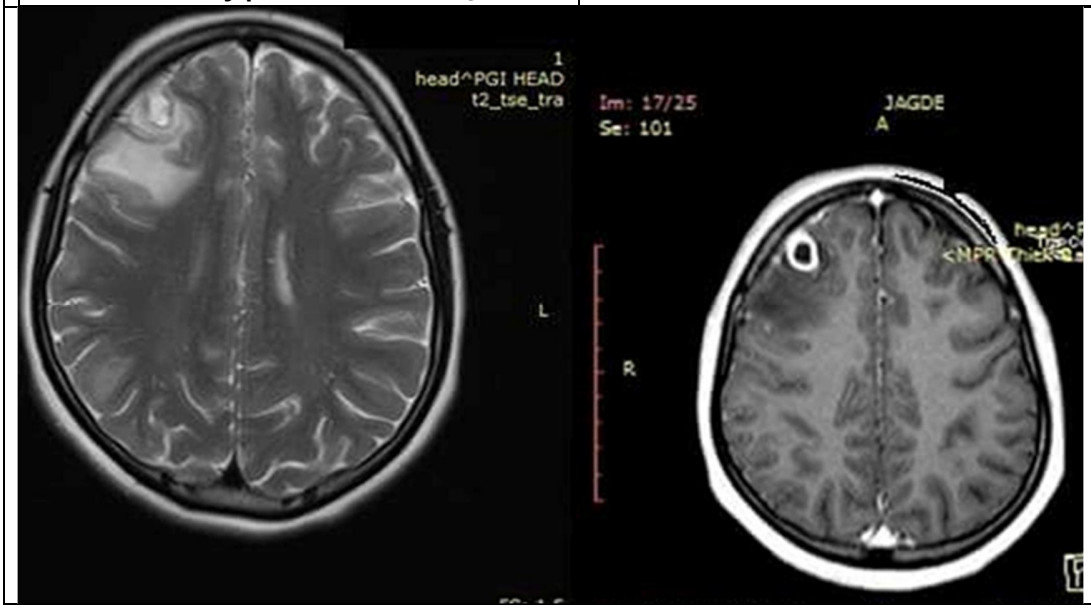
ii) Solid Caseating Tuberculoma

T2: central Hypointense signal	T1+C: Marginal / rim enhancement



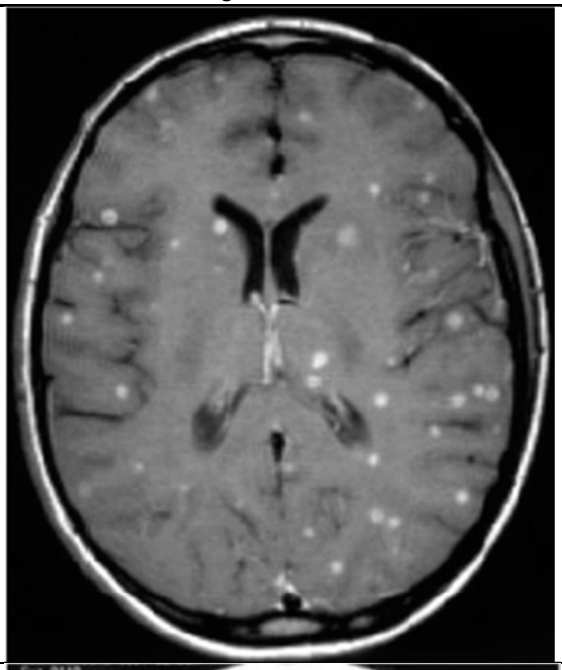
ii) Liquid Caseating Tuberculoma

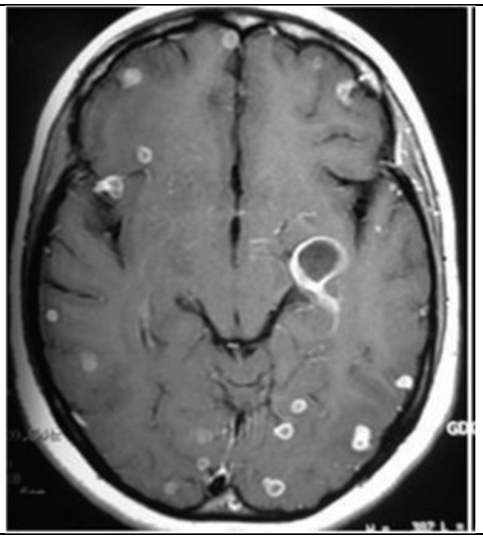
T2: central Hyperintense signal	T1+C: Marginal / rim enhancement

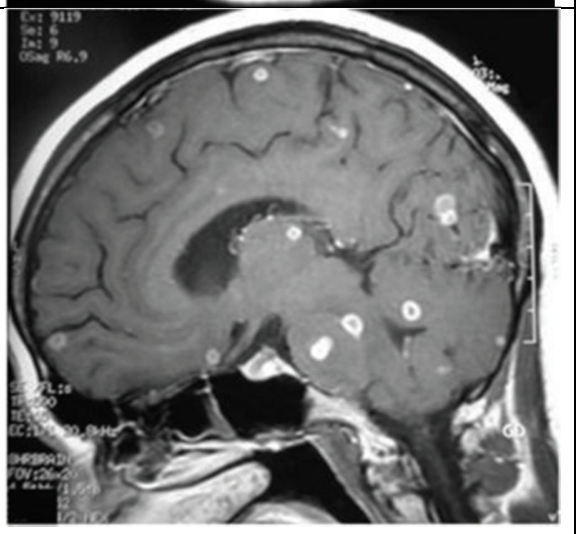


3) Miliary Brain Tuberculosis

- Numerous round, small, homogeneous enhancing lesions.



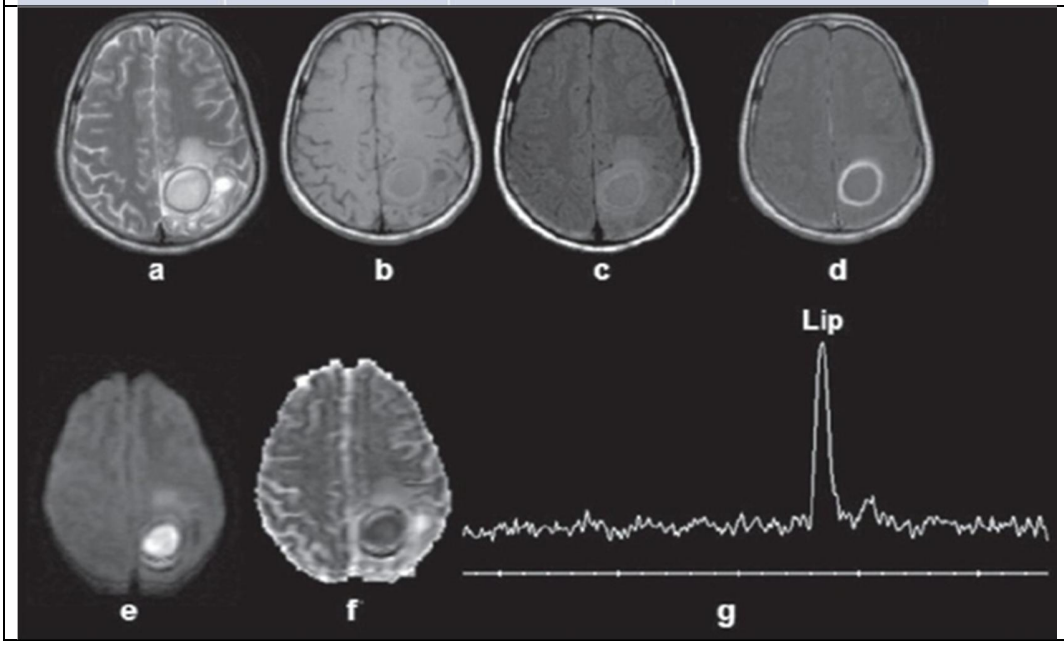




4) TB Brain Abscess

- Extremely rare.
- Solitary and larger and progress more rapidly than tuberculomas

	PYOGENIC ABSCESS	TUBERCULAR ABSCESS	FUNGAL ABSCESS
Rim	Smooth, .lobulated	Smooth, lobulated	**Crenated** with **intracavitory** projection
DWI	Restriction	Restriction	No restrctn. May show **restriction in intrcav.** Projection from wall ADC Value ....relatively lower
MRS	Acetate ,lactate,alanine, succinate,pyruvate with complex peak of **amino acid**	**Lipid p**eak	Amino acid ,lipid, lactate, multiple peaks b/w 3.6 to 3.8ppm d/t **trehalose**

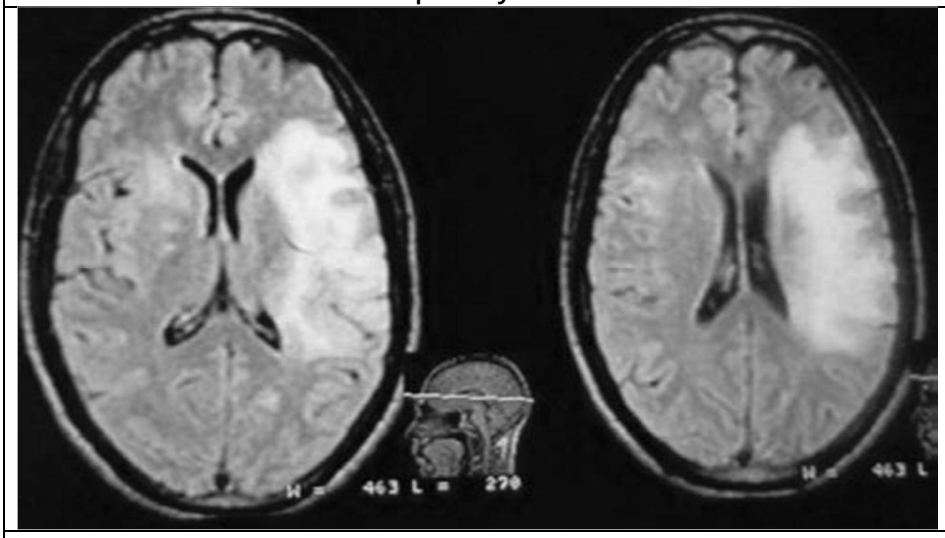


5) Tuberculous Cerebritis

- TB cerebritis is rare but has specific clinical, radiological, and pathological manifestations.
- The involved areas show extensive inflammatory exudates, Langerhans' giant cells, reactive parenchymal changes, and diffuse caseating and noncaseating microgranulomas in the cortex.

<u>Imaging findings:</u>

- T1: patchy area of hypo-intense signal
- T2: patch area of hyper-intense signal
- T1+C: small areas of patchy enhancement.



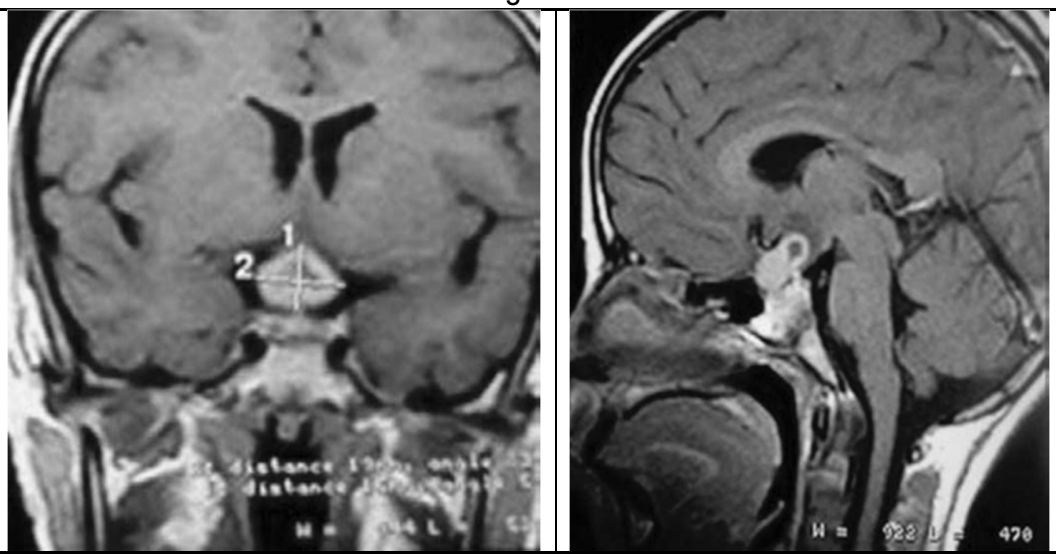
6) Tuberculous Encephalopathy

- Tuberculous encephalopathy, a syndrome exclusively present in infants and children,
- The characteristic features of this entity are the development of a diffuse cerebral disorder in the form of convulsions, stupor, and coma, without signs of meningeal irritation or focal neurological deficit.
- Pathologically, there is diffuse oedema of cerebral white matter with loss of neurons in the grey matter.
- Neuroimaging shows severe unilateral or bilateral cerebral oedema. On T2-weighted images, hyper-intensity is seen in white matter suggesting myelin loss.

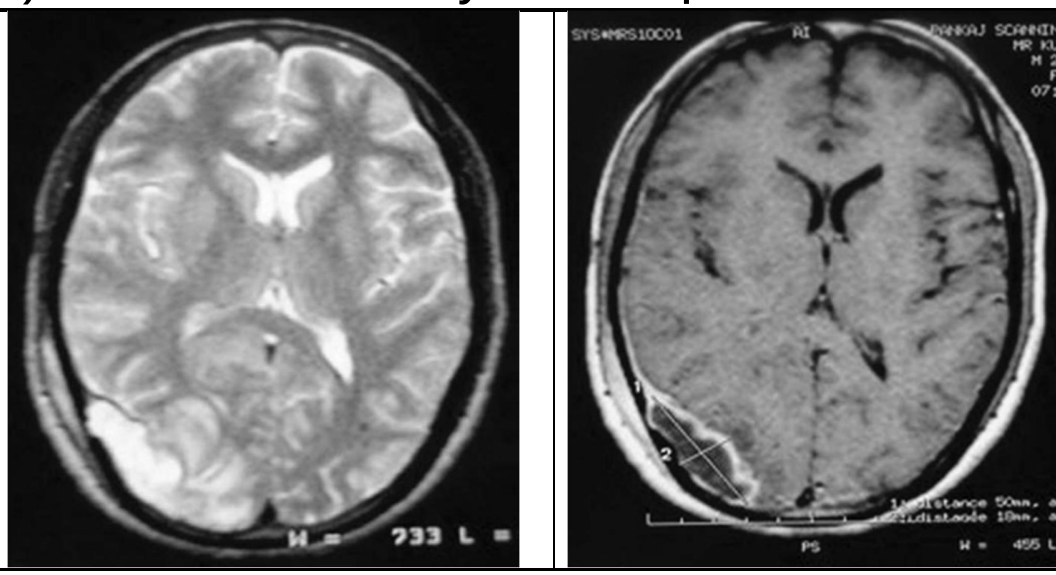
7) Hypophyseal Tuberculoma

- rare clinical entities
- Clinically simulate pituitary adenoma with or without manifestations of hypopituitarism.

- T1: isointense mass
- T2: hypointense to hyperintense
- T1+C: dense enhancement along with thickened stalk.



8) Calvarial Osteomyelitis + Epidural Abscess



9) Spinal TB

T2 hyperintense intramedullary tuberculomas	Spinal leptomeningeal TB

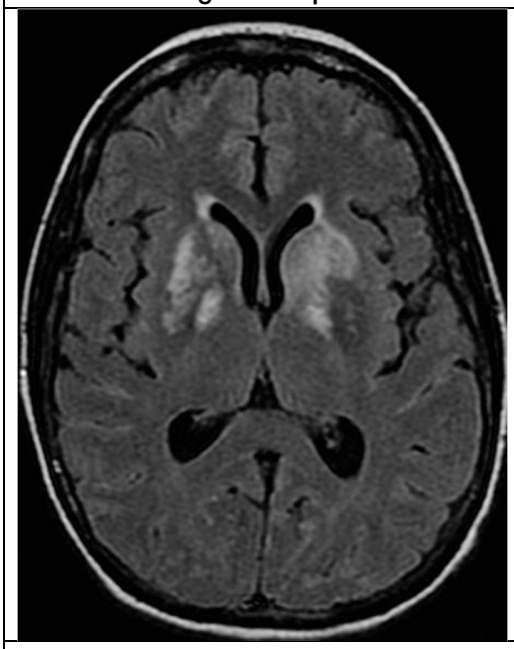
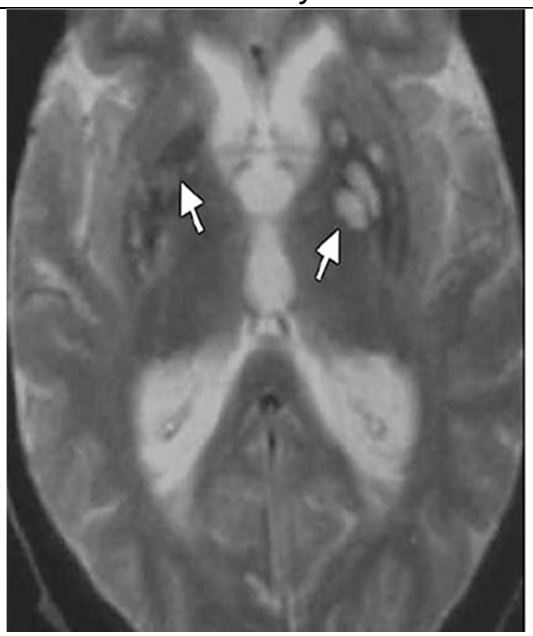
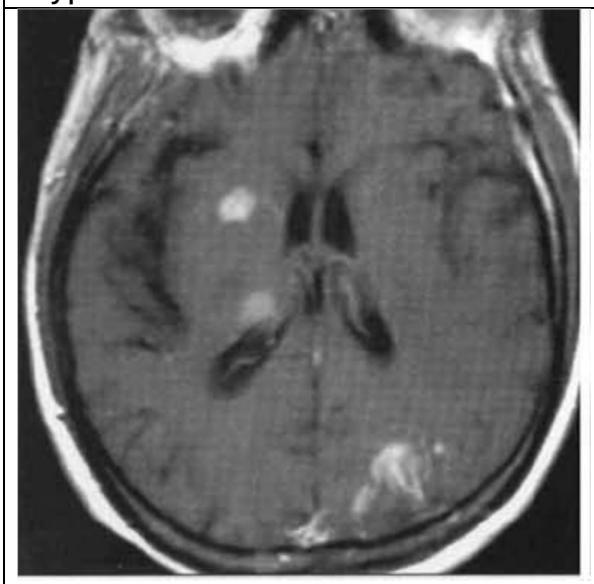
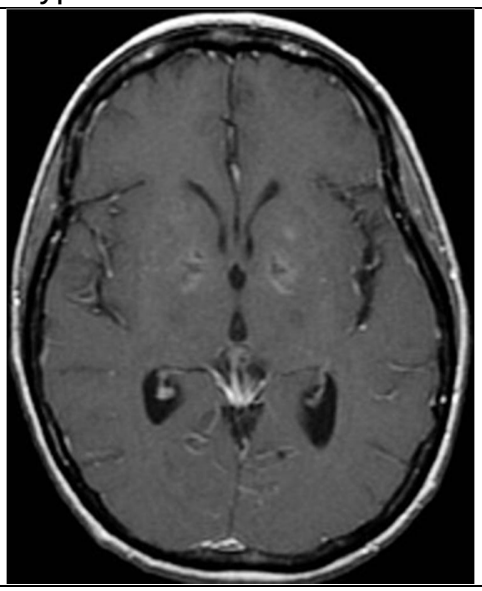
III- Fungal CNS Infection

Cryptococcosis

- is caused by Cryptococcus neoformans.
- Most common CNS fungal infection in patients with AIDS
- 3rd most common CNS infection in AIDS overall, after HIV encephalopathy and toxoplasmosis.
- AIDS patients become susceptible to cryptococcus with a CD4 count less than 100 cells/μL.

<u>Imaging findings:</u>

- Basilar Meningitis / meningioencephalitis
- Cryptococcomas:
  - T1 : low signal
  - T2 / FLAIR : high signal
  - T1 C+ (Gd) : peripheral nodular enhancement
- Gelatinous Pseudocysts:
  - Dilated basal ganglia perivascular spaces resulting from the presence of gelatinous pseudocysts
  - T1 : low signal
  - T2 / FLAIR : high signal
  - T1 C+ (Gd) : ring enhancement
- Hydrocephalus: communicating or non-communicating,

Basal meningioencephalitis	Gelatinous Pseudocysts:
	
Cryptococcomas	Cryptococcomas
	

IV- CNS Parasitic Infection

A- Neurocysticercosis

Neurocysticercosis is caused by CNS infection with the pork tapeworm Taenia solium.

Demographics:

- The disease is endemic in Central and South America, Asia and Africa.
- There is a variable time interval between point of infection and the onset of symptoms (ranging from 1 - 30 years).

Clinical presentation:

- Seizures : commonest cause of seizures in young adults in endemic areas
- Headaches
- Hydrocephalus
- Altered mental status
- Neurological deficits

Diagnosis:

- CSF serology may be helpful with the initial diagnosis especially in cases of intraventricular / subarachnoid infection.

Four Variants

- Parenchymal
- Intraventricular
- Cisternal
- Spinal

1) Parenchymal Cysticercosis

i. Vesicular stage
- Cyst with dot sign
- CSF density / intensity
- Eccentric nodule (scolex)
- No edema
- No enhancement

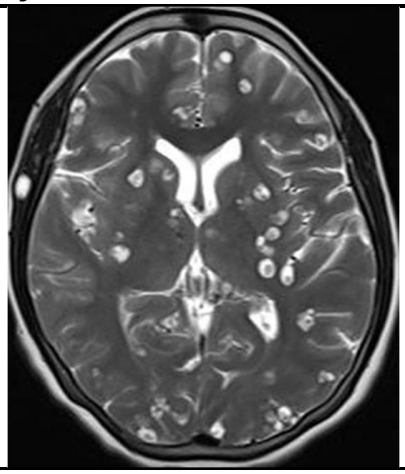
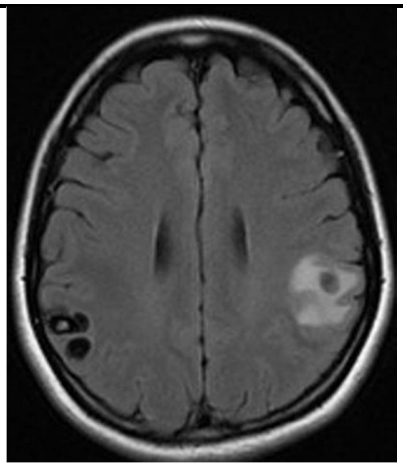
ii. Colloidal stage
- Dying scolex
- Capsule thickens
- Cyst fluid becomes turbid
- CT : hyperdense to CSF
- T1 : hyperintense to CSF
- Surrounding edema
- Marginal enhancement

iii. Granular nodular stage
- Cyst decrease in size
- Small enhancing nodules
- No edema

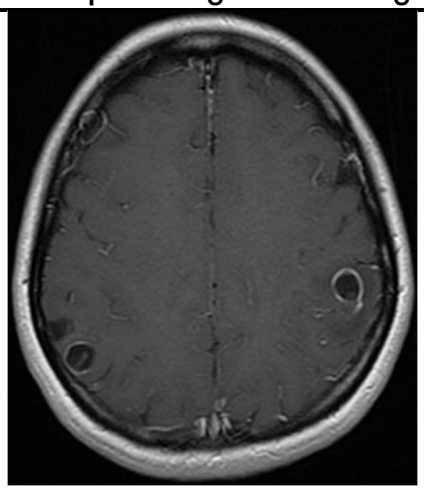
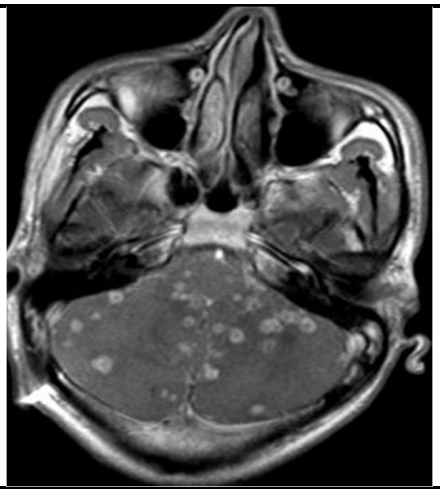
iv. Nodular calcified stage
- Cyst involutes + calcifies
- End stage quiescent calcified cyst remnant
- No oedema
- No enhancement
- signal drop out on T2 and T2* sequences
- some intrinsic high T1 signal may be present

Calcified nodules

Cyst with central dot sign

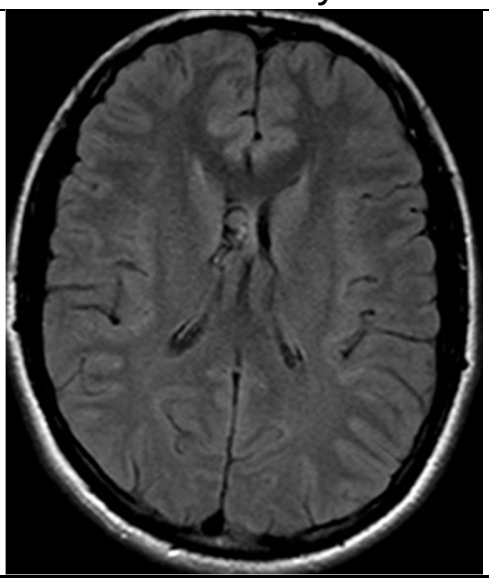



Multiple Ring Enhancing Lesions:

2) Intraventricular and Cisternal Cysticercosis

- 10-20% of neurocysticercosis
- Fourth ventricle - most common site
- Basal and sylvian cisterns
- +/- hydrocephalus
- Cyst can parallel CSF density/SI, making it difficult to detect
- Does not calcify

Intraventricular Cysticercosis	Basal Cysternal Cysticercosis
	

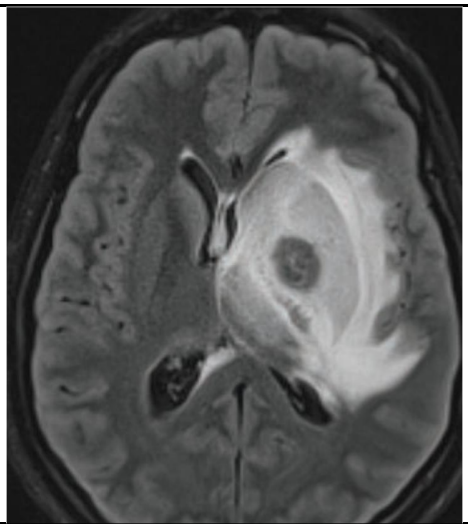
B- Toxoplasmosis

- Toxoplasmosis is the most common mass lesion in AIDS patients
- caused by the parasite Toxoplasma gondii.

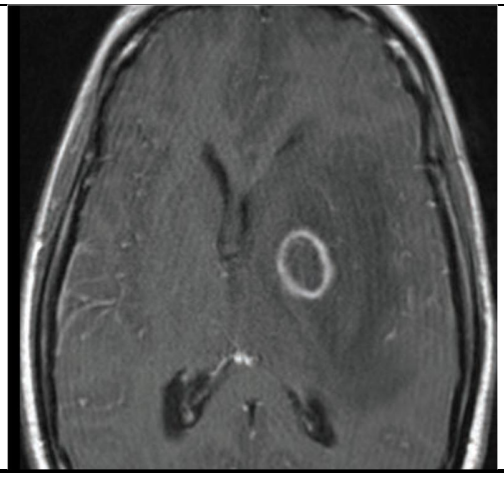
- Toxoplasmosis is the second most common CNS infection in AIDS patients, with HIV encephalitis being the most common.
- AIDS patients become susceptible to the parasite with a CD4 count less than 100 cells/μL.

<u>Axial T2/FLAIR:</u>

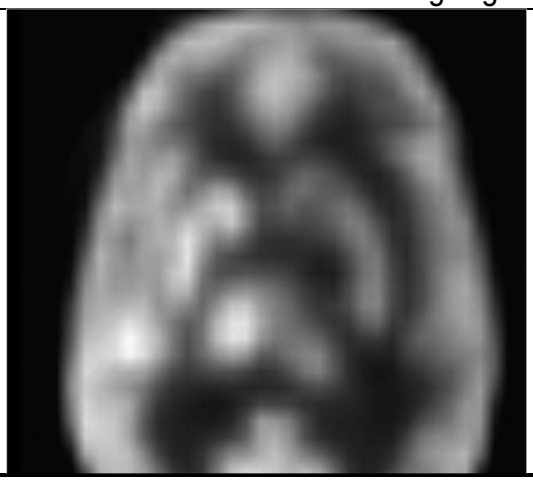
- A rounded hypo- to isointense lesion in the left basal ganglia surrounded by hyperintense signal extensive edema



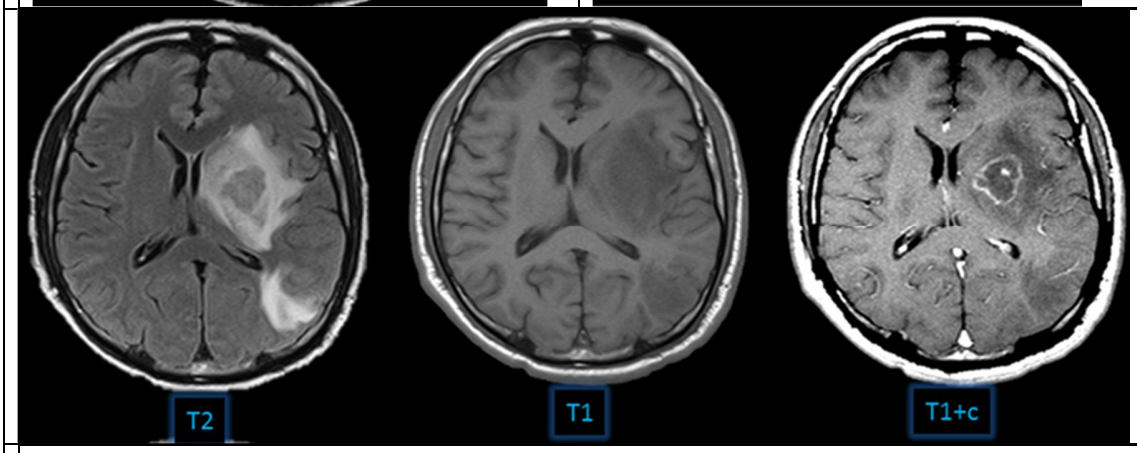
T1+C: Ring-enhancing lesions in the basal ganglia.



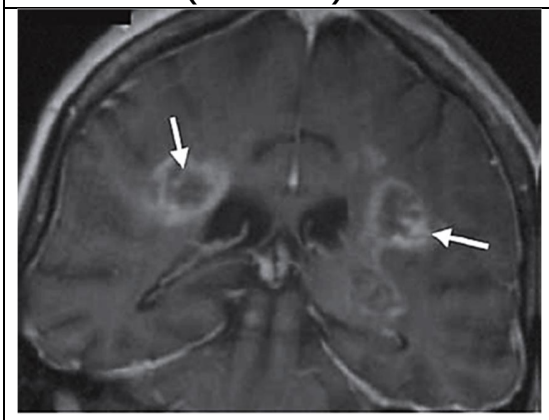
F-18-FDG PET scan shows asymmetrical, relatively decreased metabolism of the left basal ganglia

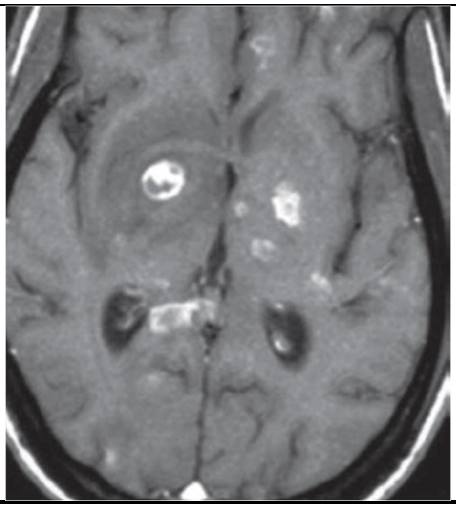


NCCT	CECT



"Target Sign" = Ring-enhancing lesions with eccentric nodules (arrows).





V- CNS Viral Infection

A- Herpes Encephalitis

- is one of the most common viral infections.
- Caused by reactivation of latent HSV-1 within the trigeminal ganglion
- Infants => HSV type 2 (Genital)
- Children and adults => HSV type 1 (oral)
- 50%-70% mortality

Mechanism of spread:

- Axonal spread from reactivation in trigeminal ganglion

- Clinical picture
  - Fever, headache, seizures
  - Acute altered mental status
  - PCR of CSF most useful for diagnosis

Location:

- Bilateral disease, but asymmetric
- Limbic system:
  - Medial temporal lobes
  - Inferior frontal lobes
  - Insula
  - Cingulate gyri typical
- Basal ganglia usually spared

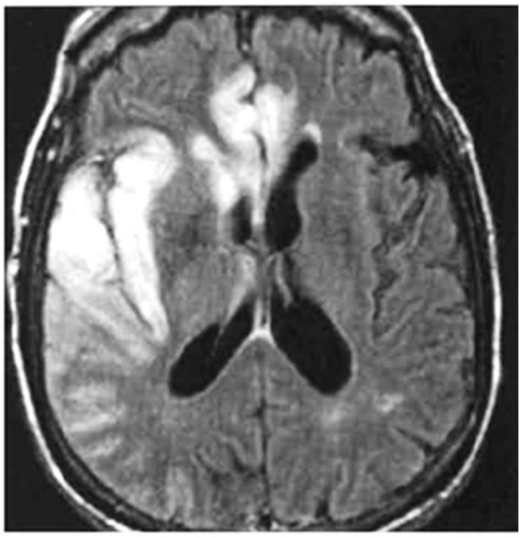
MRI findings:

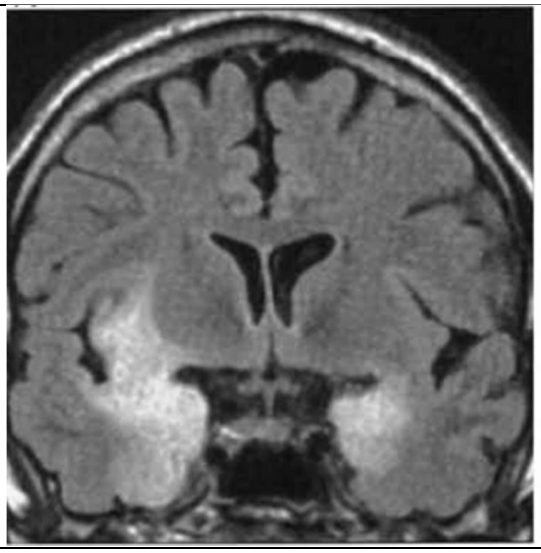
- T2/FLAIR: hyperintense lesions
- T1: +/- petechial hemorrhage are characteristic findings.
- T1+C: Patchy or gyriform enhancement of temporal lobes (late feature).
- DWI: Restricted Diffusion = DWI high signal + decreased ADC values.

MRS findings:

- Elevated peaks of lactate, choline, and myoinositol
- Reduced NAA signal

T2/FLAIR: hyperintense signal of medial temporal + Inferior frontal + Insular





Restricted DWI

Gyral enhancement

B- Flavivirus Encephalitis

Flavivirus infections such as

Japanese encephalitis
West Nile fever
Murray Valley fever

Geographic Distributions:

- Japanese encephalitis being common in Asia.
- West Nile fever in the Middle East (now also in North America)
- Murray Valley fever in Australia.

Clinical Manifestations:

- Prodromal phase: of fever, rigors, headache, rashes, and body aches
- *followed by*
- CNS symptoms: include dyskinesia, dystonia, tremors, drooling, dysarthria, altered consciousness, seizures, and coma

Definitive serologic diagnosis:

- Detection of antibodies in serum and CSF at enzyme-linked immunosorbent assay

Location:

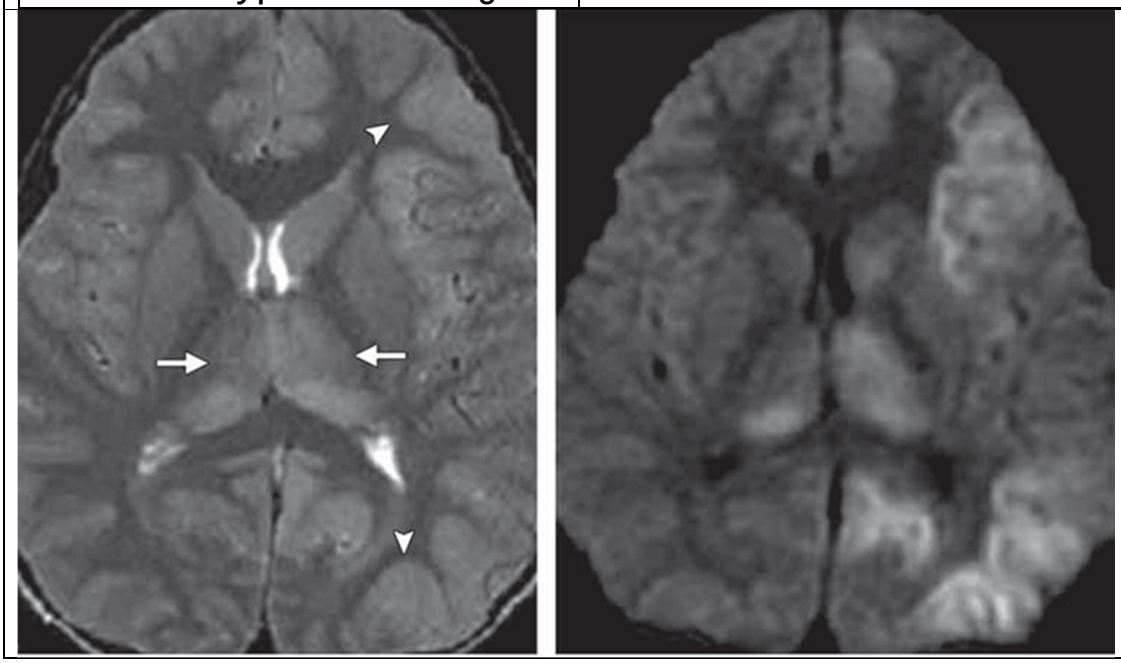
- Symmetric involvement of the deep gray matter structures.
    - Posteromedial Thalamus
    - Basal ganglia, Substantia nigra
    - Red nucleus, pons
    - Hippocampi
- Cerebral cortex
- Cerebellum.

MRI findings:

- T2/ FLAIR: hyperintensity
- T1: +/- Intralesional hemorrhages
- DWI: Restricted diffusion have also been described

Bilateral Thalami + BG + Cortex

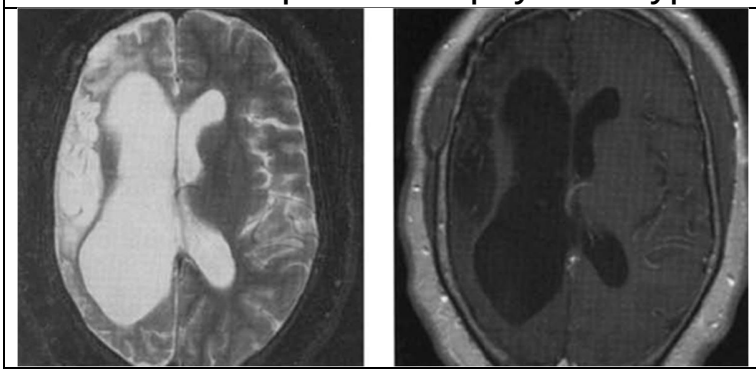
T2/FLAIR: Hyperintense signal	DWI: Restricted



C- Rasmussen Encephalitis

- Rare childhood syndrome
- Chronic progressive unilateral inflammation of the brain of uncertain cause.
- Characterized by medically refractory focal seizures and progressive multifocal neurologic symptoms such as hemiplegia and cognitive impairment.

Cerebral Hemisphere Atrophy + T2 Hyperintensity



D- Acquired CMV Infection

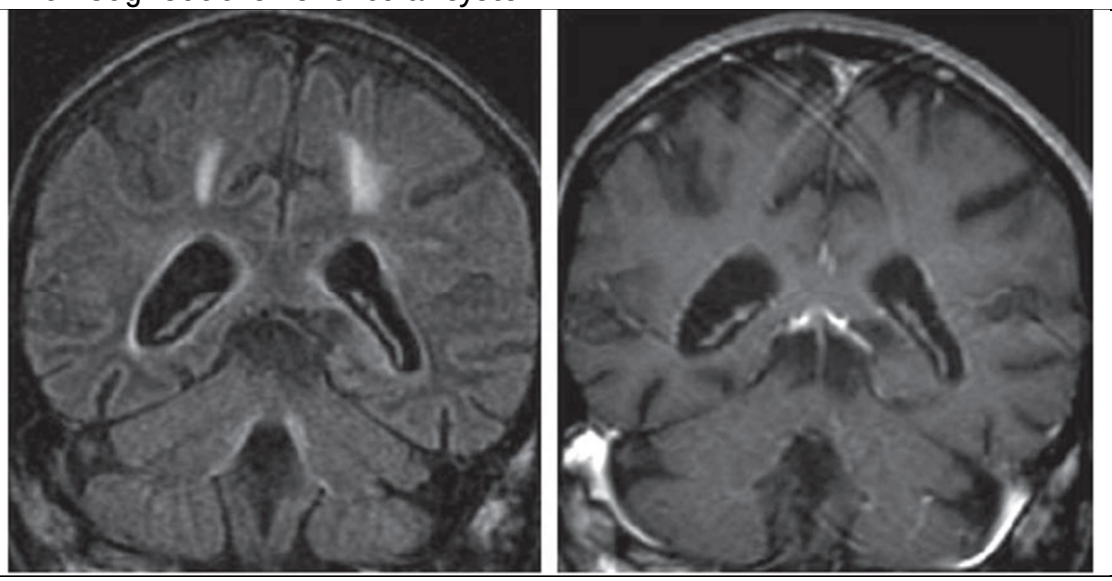
- Cytomegalovirus (CMV) encephalitis only affects the immunosuppressed, typically when the CD4 cell count < 50 cells /μL.

CNS involvement forms:

- C => Encephalitis
- M => Meningitis / Meningoencephalitis
- V => Ventriculitis = Ependymitis
- Myelitis,
- Polyradiculitis
- Retinitis

Imaging findings:

- Ventriculitis = Subependymal FLAIR hyperintensity and enhancement throughout the ventricular system.



VI- HIV

- The human immunodeficiency virus (HIV) is a retrovirus that infects cells of the immune system and destroys or disrupts their function

Neurologic Manifestations of AIDS patients (10):

- HIV infection itself:
  - AIDS Dementia Complex = HIV Encephalopathy
  - Progressive Multifocal Leukoencephalopathy (PML)
- Secondary opportunistic infections: (discussed before)
  - Parasite => Toxoplosmosis
  - Viral => CMV
  - Fungal => Cryptococcosis
  - Bacterial => Staph / Strept
  - Granulomatous => TB
  - Neurosyphilis
- Neoplasms
  - Primary CNS Lymphoma
- Drug-related complications of therapy
  - Immune Reconstitution Inflammatory Syndrome (IRIS)

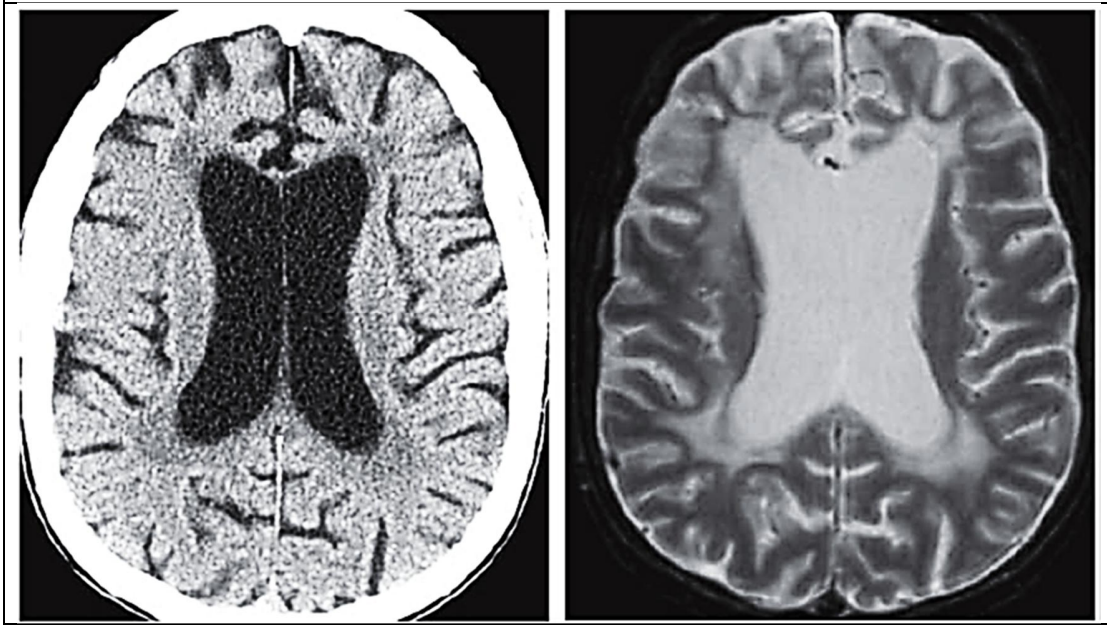
1) AIDS Dementia Complex
HIV Encephalopathy / Encephalitis

Neurodegenerative syndrome characterized by progressive

- Cognitive and motor impairment.
- Atrophic changes involving the brain.
- White matter demyelination and gliosis.

Imaging findings:

- Cerebral atrophy +
- T2/ FLAIR bilateral and symmetrical hyperintense signal of the periventricular and deep WM
- Spares the subcortical U-fibers and tends to be symmetric
- No enhancement
- No restricted DWI



2) Progressive Multifocal Leukoencephalopathy (PML)

- is a progressive demyelinating disorder that results from a viral infection of the myelin producing oligodendrocytes.
- The infecting agent is the John Cunningham virus, which is a DNA papovavirus.

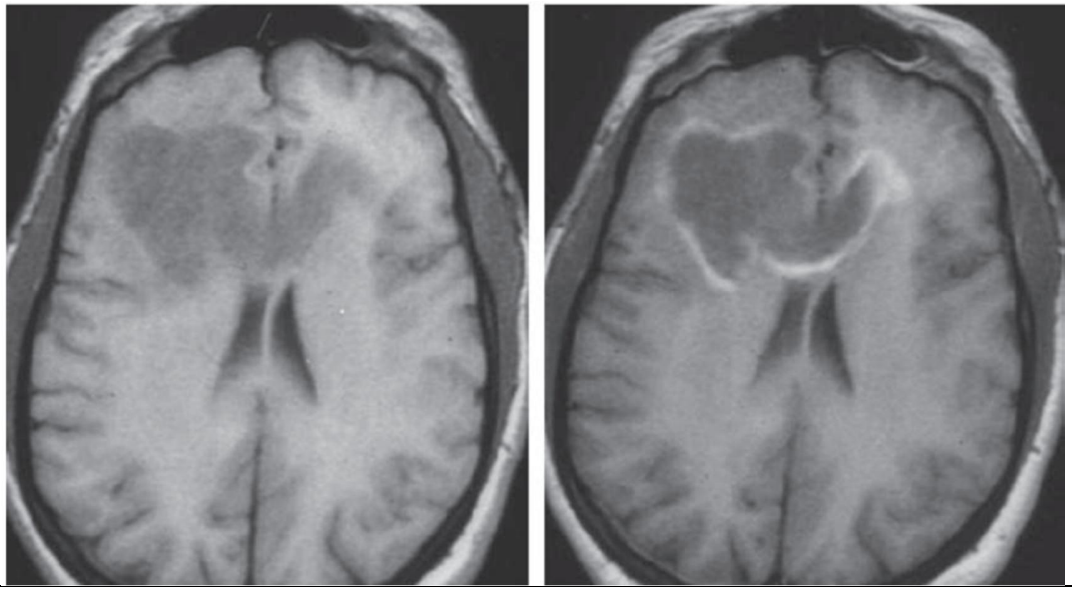
Clinical picture:

- Progressive neurologic decline and patients develop cognitive impairment, altered mental status, and personality changes.
- Motor and sensory changes also occur, and the patients may develop seizures.

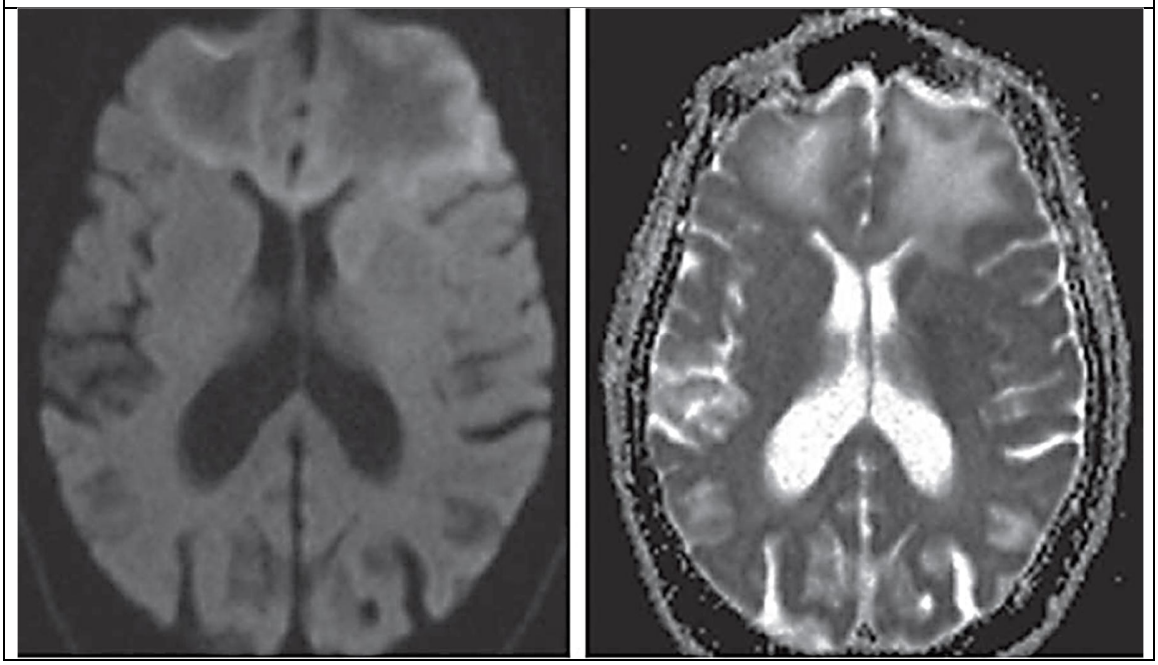
Imaging findings:

- Bilateral multifocal asymmetrical
- Involve periventricular and subcortical WM.
- Mass effect and hemorrhage are unusual.

T1+C: NO enhancement, but faint peripheral enhancement has been described => better prognosis



Peripheral restricted diffusion
Central facilitated diffusion



HIV Encephalopathy	PML
Bi & symmetrical	Uni or Bi & Asymmetrical
Periventricular and deep WM	Peripheral subcortical WM
No mass effect	Mild mass effect
No hemorrhage	+/- Hemorrhage
No enhancement	No or peripheral enhancement
No restriction	No or peripheral restriction

3) Primary CNS Lymphoma

- Primary CNS lymphoma (PCNSL) are uncommon tumours, accounting for only 1% of malignant CNS tumours.
- Primary = No co-existing systemic disease at the time of diagnosis.
- Strong association with HIV/AIDS and other immunocompromised

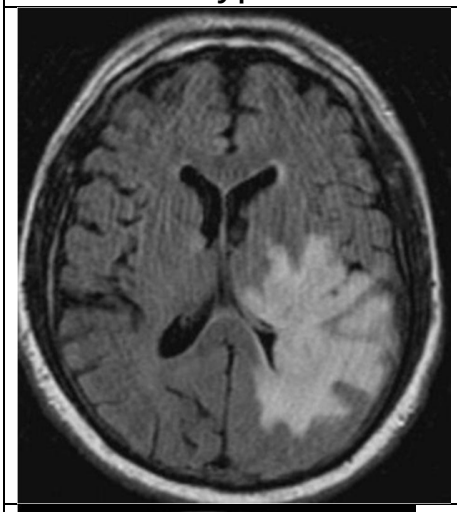
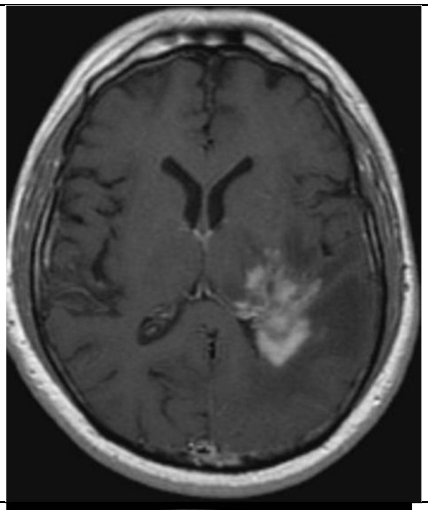
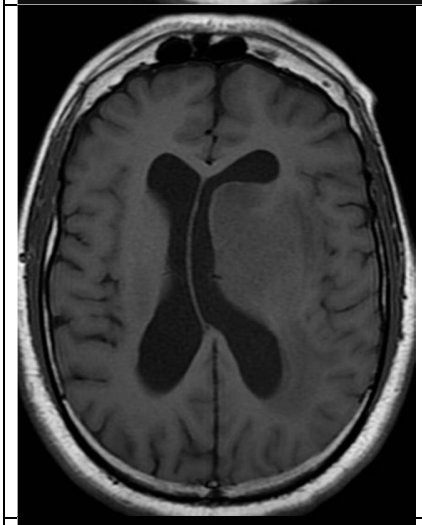
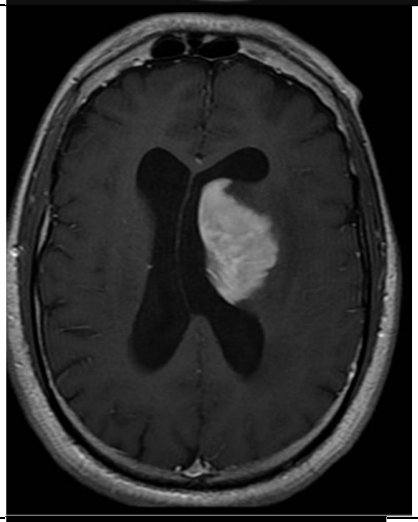
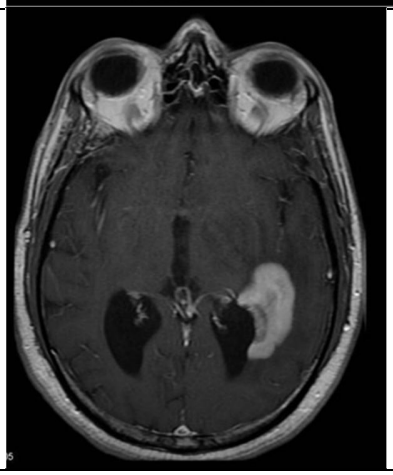
Location:

- Supratentorial (75-85%)
- Subarachnoid / Ependymal surfaces.
- Crossing the corpus callosum
- Multiple basal ganglia lesions (DD: Toxoplasmosis)

Imaging findings:

- NCCT: hyperattenuating + No hemorrhage
- T1: hypointense
- T2: hyperintense
- +C: Moderate/Strong homogenous enhancement.
- Little mass effect
- Limited surrounding vasogenic oedema.

- MRS: large choline peak
- MR Perfusion: Moderate Increased rCBV
- Scintigraphy Thallium 201/PETCT: shows increased uptake

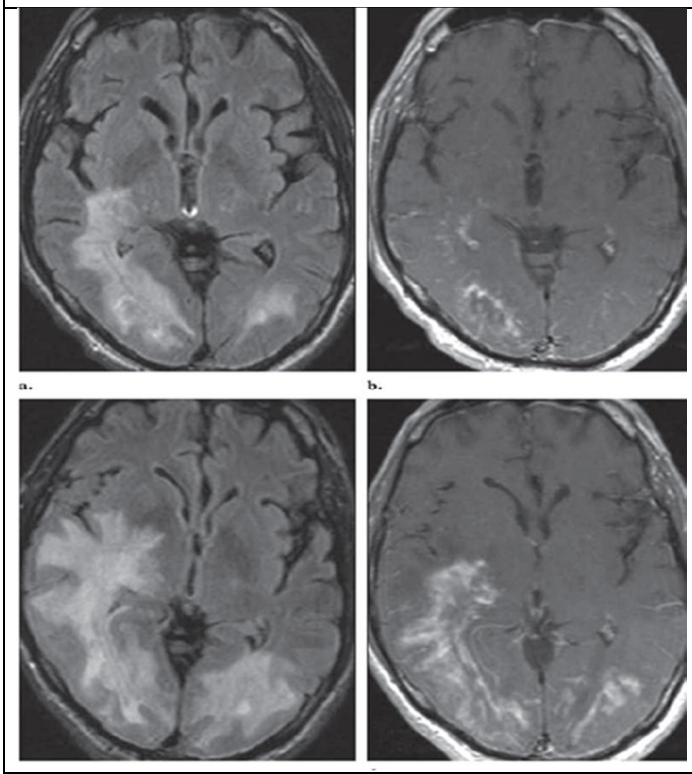
T2/FLAIR: hyperintense	T1+C: Moderate enhancement.
	
	
	

Toxoplasmosis	Lymphoma
Basal ganaglia	Subependymal spread
Single or multiple lesions	Single lesion
+/- Hemorrhage	No hemorrhage
Ring or nodular enhancement	Solid enhancement
Follow-up evaluation after adequate treatment (multidrug regimen of pyrimethamine sulfadiazine, and folic acid)	
Regressive course 2-4w The brain may appear normal or encephalomalacia or calcification	Lack of resolution suggest lymphoma
Thallium-201 SPECT or FDG PET	
Negative	Positive
MRI perfusion	
Decrease in cerebral blood volume (rCBV)	Increased rCBV
MRS	
Elevated peaks of lipid and lactate without elevated choline levels	Elevated peaks of choline

4) Immune Reconstitution Inflammatory Syndrome (IRIS)

- HAART succeeds in suppressing HIV replication and improving cellular immunity, which protects HIV-infected patients against opportunistic infections
- However, in a few of these patients, partial restoration of specific immunity may worsen a preexisting disease; the resulting condition is referred to as immune reconstitution inflammatory syndrome (IRIS)
- IRIS is not caused by a relapse or recurrence of the preexisting disease, and its exact etiology is unknown.

- Patients with IRIS demonstrate paradoxic deterioration in their clinical status when their CD4 counts rise and viral replication appears to be under control and death from IRIS has been reported (17).
- IRIS occurs in the initial months after the onset of HAART.
- The neuroimaging findings vary, depending on the underlying pathologic conditions, and may be atypical, such as prominent, progressive enhancement and mass effect seen in PML.



VII- TORCH

A- Congenital CMV Infection

- Congenital CMV infection is one of the most common congenital viral infections in the world.

CNS involvement forms:

C => Calcification + Cysts (Periventricular)

M => Migration disorder + WM Delayed Myelination + WM Lesions

V => Ventriculomegaly + Brain atrophy + Microcephaly

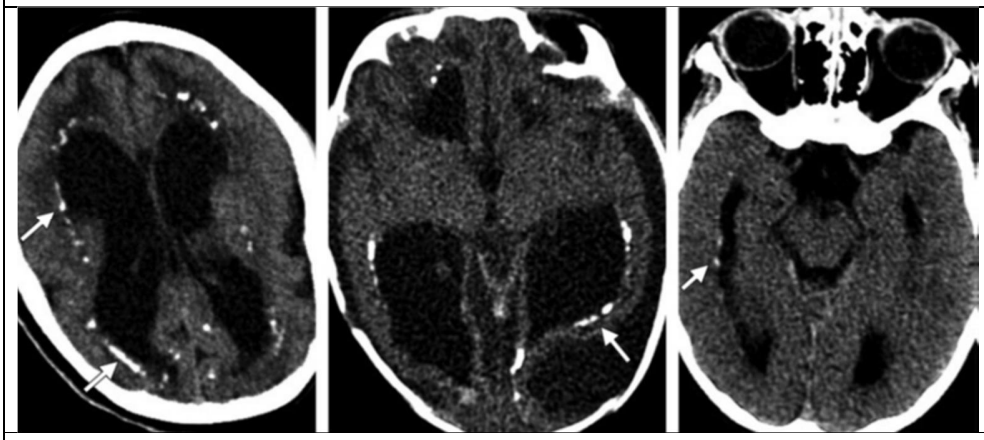
1) Intracranial Calcifications:

Location: Periventricular – basal ganglia

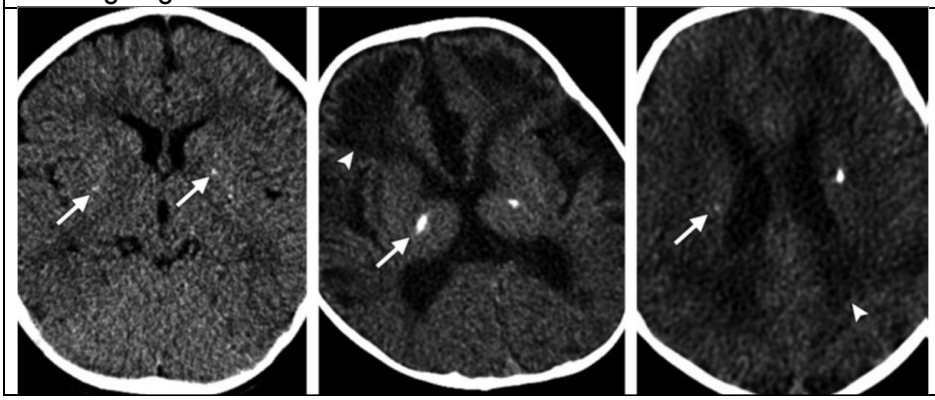
Shape:

- Thick and chunky
- Faint and punctate

Ventricular dilatation + Periventricular calcification

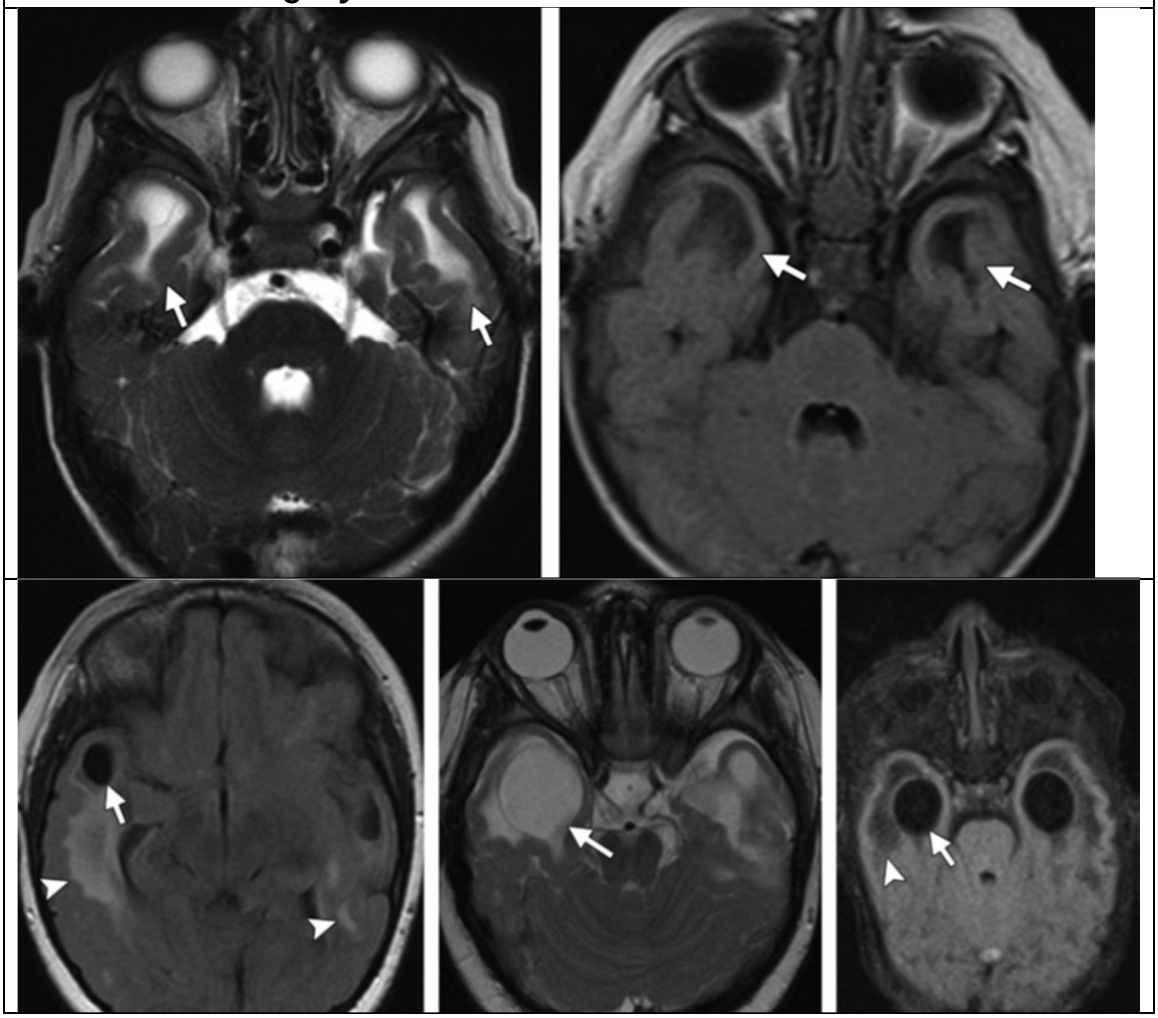


Basal ganglia calcification + WM lesions



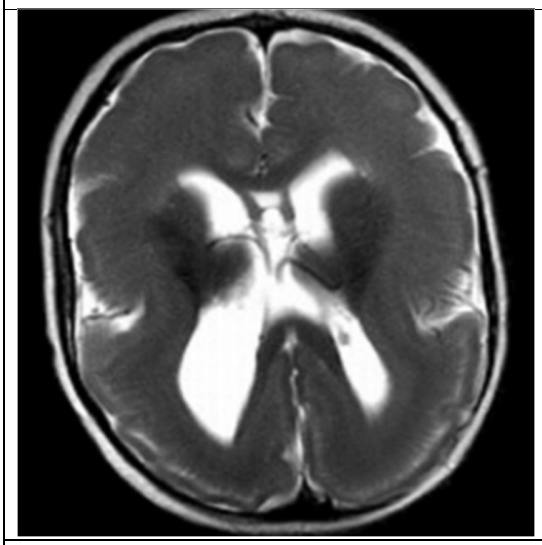
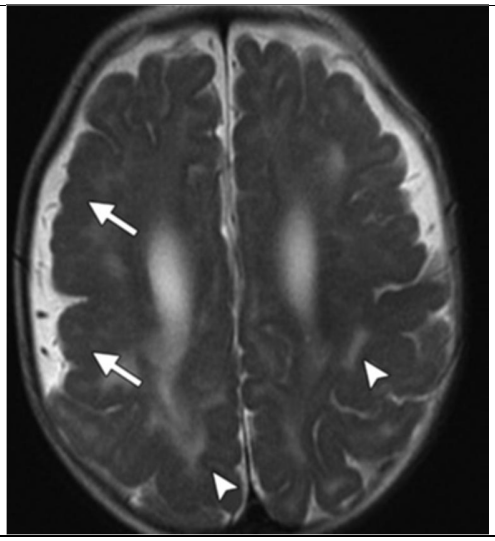
2) Periventricular Cysts

Anterior Temporal cysts / Vacuolization + Ventriculomegaly + WM lesions

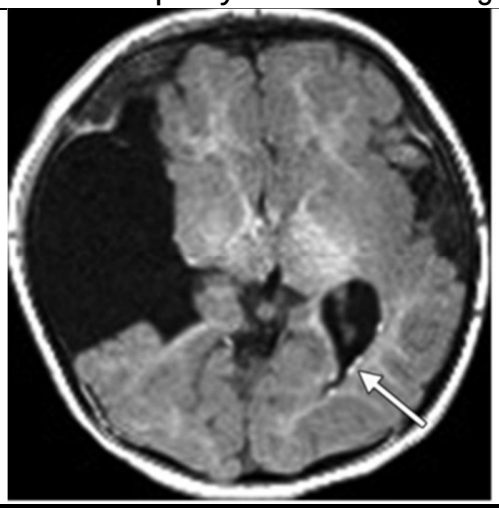


3) Migrational Abnormalities

- Lissencephaly, pachygyria, diffuse or focal polymicrogyria are the most common migrational abnormalities

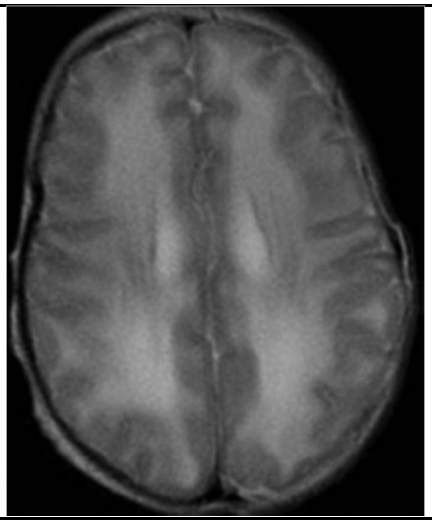
Lissencephaly + Ventriculomegaly + WM lesions	Polymicrogyria + Ventriculomegaly + WM lesions
	

Schizencephaly + Ventriculomegaly + periventricular Calcifications



4) Delayed Myelination

- Delayed myelination is nonspecific and may be seen in a variety of conditions.
- Disturbed myelination is more common in congenital CMV infection than generalized delayed or decreased myelination



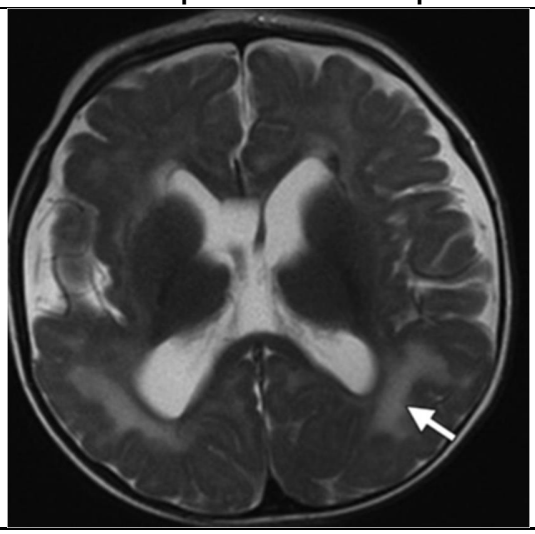
5) WM Lesions

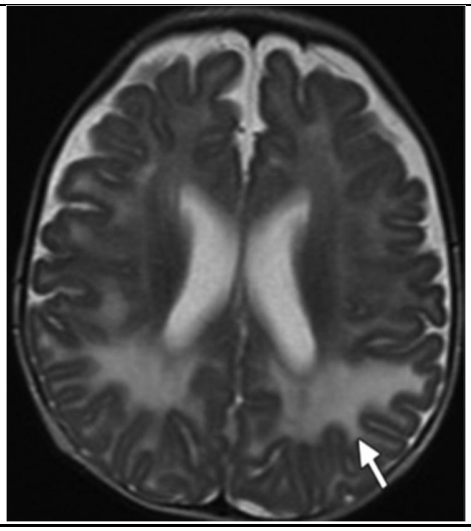
Patterns:

- Focal posterior predominant (parietal & occipital)
- Focal frontal-predominant
- Diffuse
- Random pattern

- T2/FALI R: Hyperintense signal

Posterior-predominant pattern of WM involvement

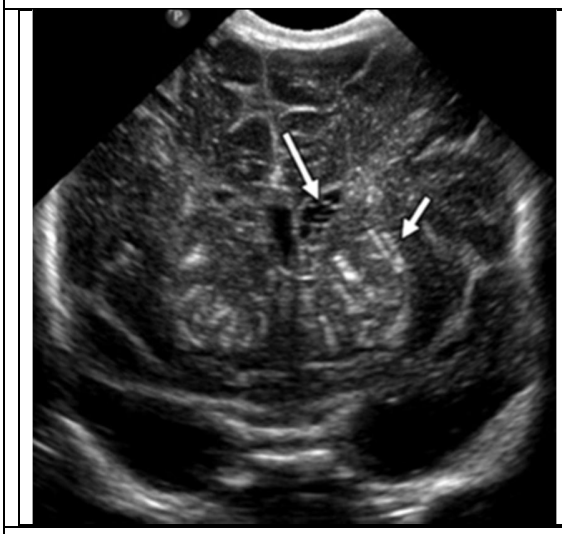


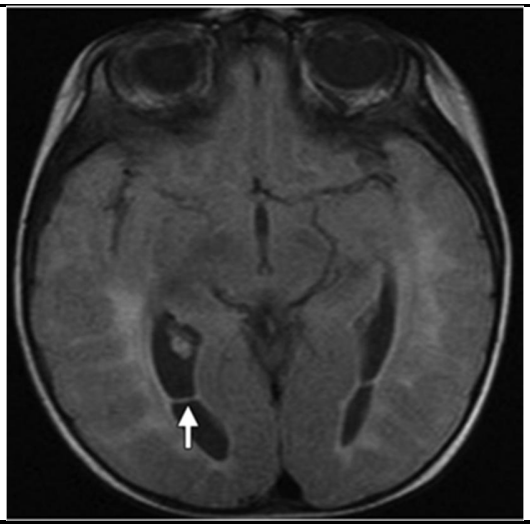


6) Ventriculomegaly+ Cerebral Atrophy + Microcephaly

- 2nd most common finding of congenital CMV infection.

7) Ventriculomegaly + Ventricular Adhesions





8) Lenticulostriate Vasculopathy

- Unilateral or bilateral curvilinear echogenic streaks within the basal ganglia and thalami
- Likely represents a mineralizing vasculopathy of the lenticulostriate vessels.
- A nonspecific finding

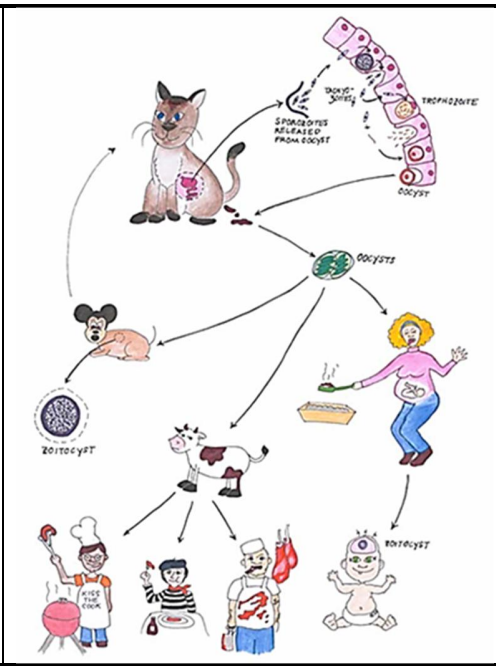
B- Toxoplasmosis

Causative organism:

- Toxoplasma gondii

Human transmition:

- Ingestion of sporulated oocysts (cat feces + incubation)
- Ingestion of zoites (undercooked meat)
- Congenital infection (only during acute stage)
- Organ transplants
- Chronic infection in donor
- Immunosuppression
- Blood transfusions (only during acute stage)



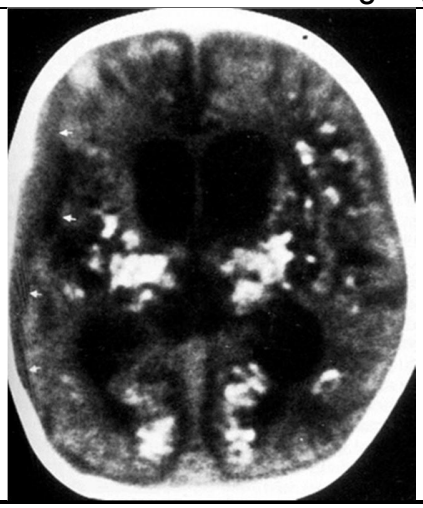
Acute infection during pregnancy to fetus

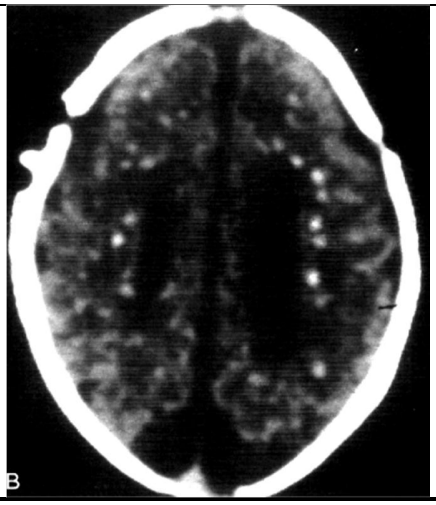
→ 1st trimester 17% Abortion
→ 2nd trimester 25% Fetal death
→ 3rd trimester 65% Sub clinical infection

CNS manifestations:

- Hydrocephalus + Microcephaly + Calcifications

Calcifications: Basal ganglia + Periventricular + Cortical





C- Herpes Simplex Encephalitis

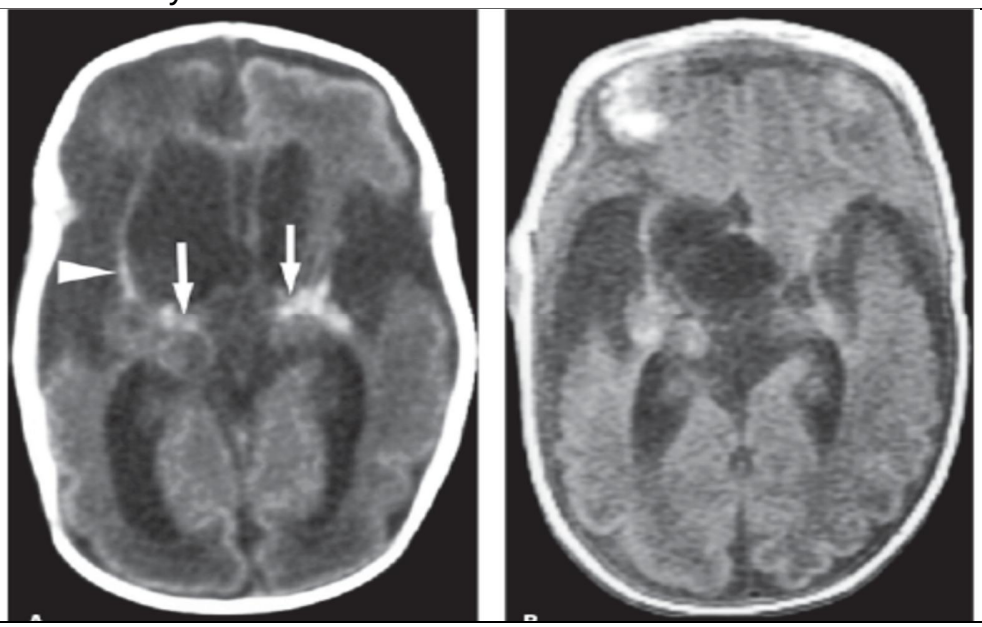
- Infants => HSV type 2 (Genital)
- Children and adults => HSV type 1 (Oral)

HSV type 2 infection

- is a sexually transmitted disease and is associated with genital lesions.
- The congenital infection is usually transmitted at birth from contact with infected secretions.
- CNS involvement is seen in 30% of these children
- 80% do not survive the infection.
- True congenital infection acquired in utero is rare.

CNS manifestations:

- Microcephaly + Microphthalmos + Ventriculomegaly
- Multicystic encephalomalacia
- Parenchymal calcification



D- Rubella

- Rare
- Ocular (cataract , glucoma)
- Cardiac (PDA , Pulmonic stenosis)
- Hearing problem.

CNS manifestations:

- Atrophy
- Microcepalia
- Calcifications: Periventricular, BG, cortical.
- Delayed Myelination
- CT petrous : Congenital inner ear abnormalities

VIII- Creutzfeldt-Jakob Disease

- CJD is a transmissible fatal neurodegenerative disorder caused by prions (self-replicating proteinaceous infectious particles).

- Patients with CJD typically present with rapidly progressive dementia, myoclonus, and multifocal neurologic dysfunction.

4 Subtypes:

- Sporadic
- Familial
- Iatrogenic
- Variant CJD: associated with bovine spongiform encephalopathy ("mad cow disease"), transmitted through consumption of bovine spongiform encephalopathy–contaminated meat.

Clinical Diagnosis:

- Brain biopsy or autopsy: is required for a definitive diagnosis
- EEG: Generalized periodic sharp wave complexes
- CSF: detection of 14-3-3 proteins in the CSF

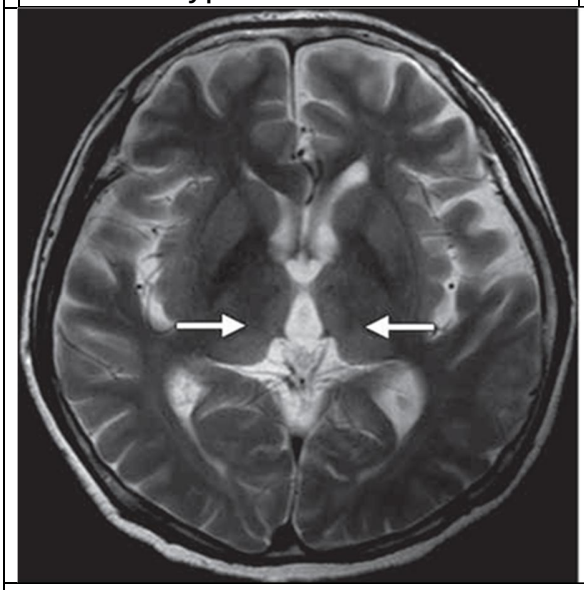
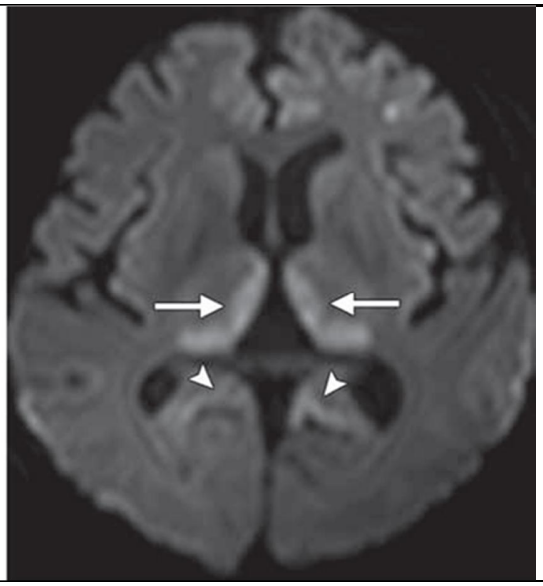
Location:

- Basal ganglia (Caudate & Putamen > GB)
- Thalamus "pulvinar sign" or hockey stick sign.
- Cerebral cortex (Cortical ribboning)

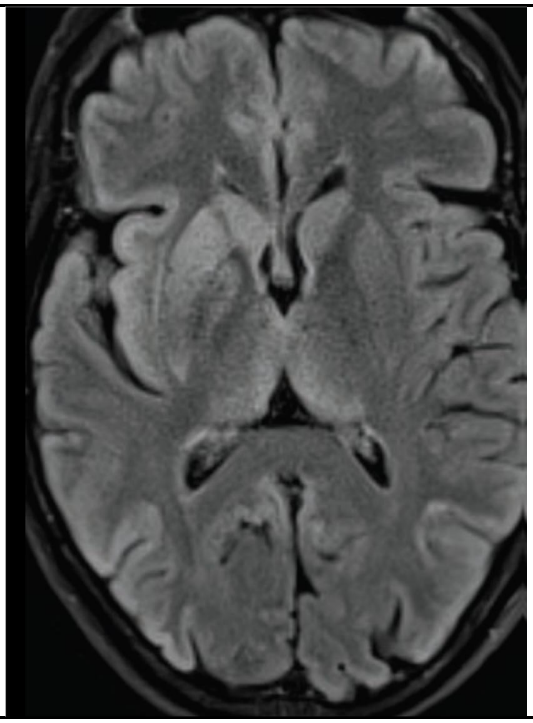
Imaging findings:

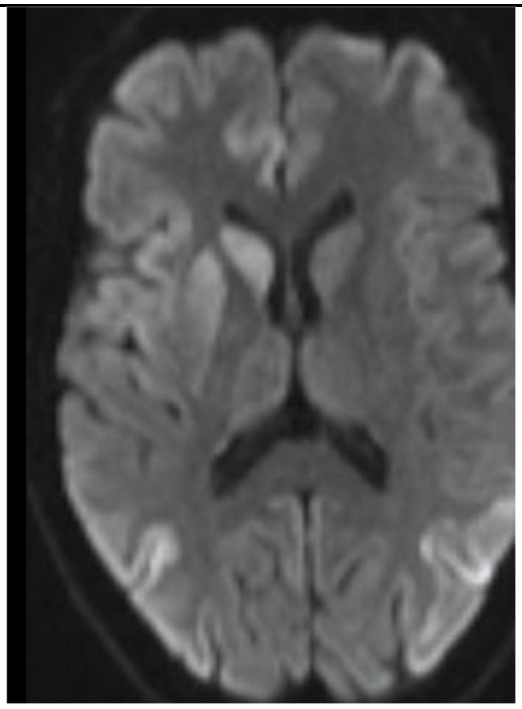
- T2 & FLAIR: Progressive hyperintensity
- DWI: Restricted
- +C: NO enhancement.

Bilateral thalamic “Pulvinar sign” or “hockey stick sign”

Axial T2: Hyperintense	Axial DWI:
	

Bilateral BG + Cerebral cortex + Thalami





Demyelinating WM Diseases

❖ Primary / Idiopathic:
- Multiple sclerosis = MS

❖ Infectious WM disease:
- Acute disseminated encephalomyelitis (ADEM)
- Hurst's disease = Acute hemorrhagic leukoencephalitis
- Progressive multifocal leukoencephalopathy (PML)
- HIV-associated encephalitis (AIDS)
- Subacute sclerosing panencephalitis
- Lyme disease

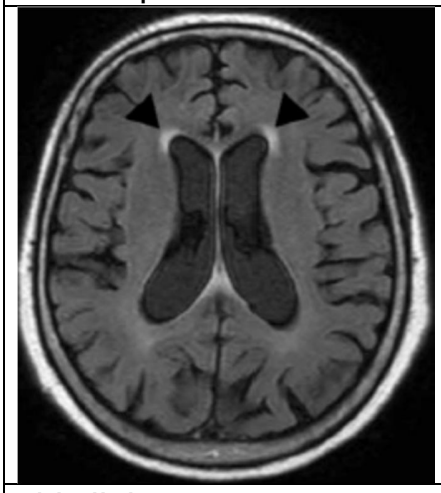
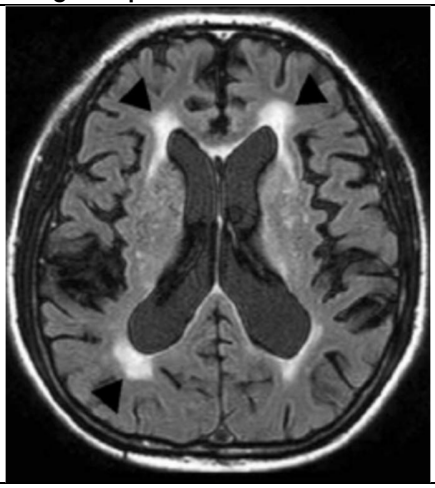
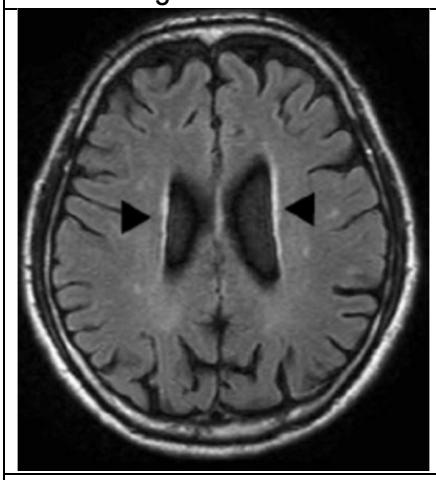
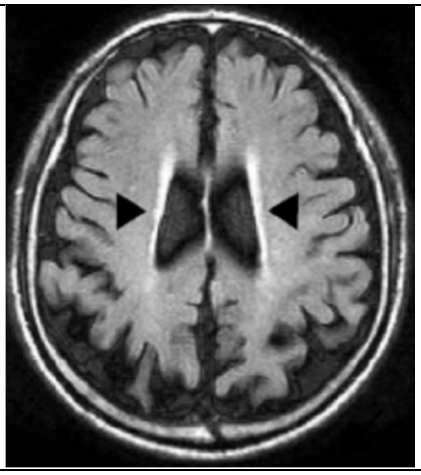
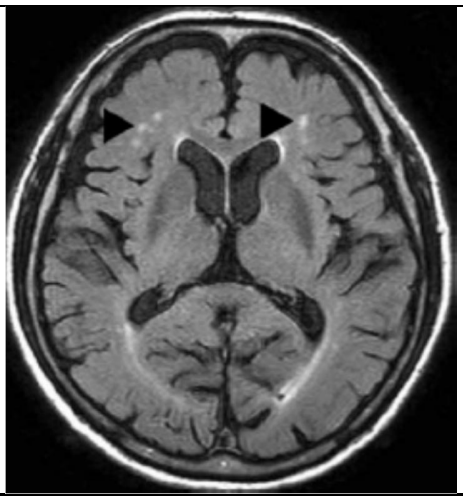
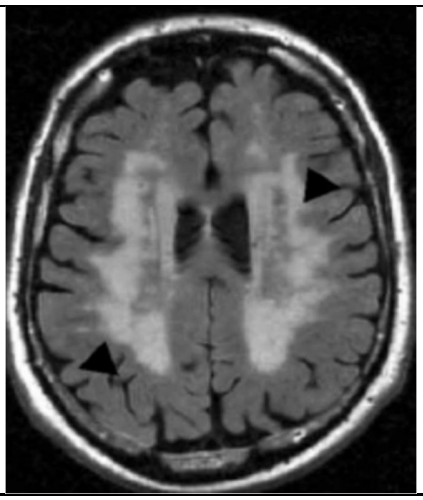
❖ Metabolic / Nutritional:
- Osmotic Demyelination Syndrome
- Alcohol
  - Marchiafava-Bignami Disease (MBD)
  - Wernicke Encephalopathy
- B12 Deficiency
- Hypothyroidism
- Hypoglycemia
- Nonketotic Hyperglycemia
- Hepatic Encephalopathy
- Acute Hyperammonemia / Acute hepatic failure

❖ Vascular WM Disease
- Chronic Hypertensive (Binswagner) = Subcortical Arteriosclerotic Encephalopathy (SAE)
- Acute Hypertensive (PRESS)
- Cerebral autosomal dominant arteriopathy with subcortical infarction & leukoencephalopathy (CADASIL)
- Vasculitis

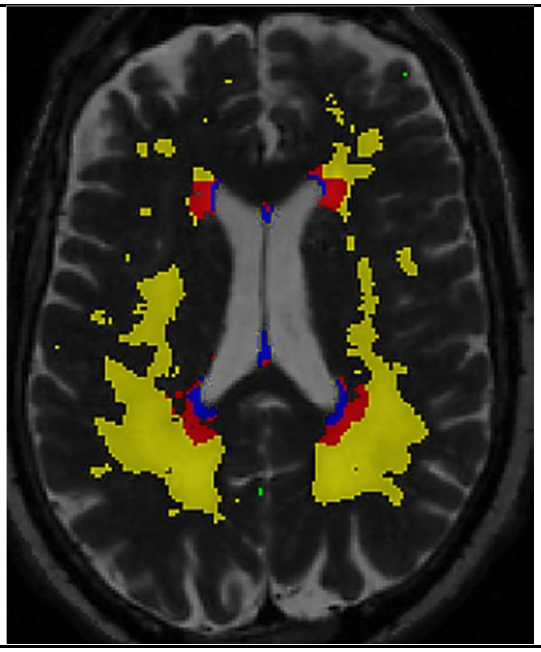
❖ Toxic Poisoning:
- Cyanide / Methanol / Carbon monoxide
- Toxic Leucoencephalopathy

❖ Iatrogenic WM Diseases:
- Drug abuse
- Post Radiotherapy
- Post Chemotherapy

Patterns of WM Lesions

Small cap	Large cap
	
Thin lining	
	
Punctate WM foci	Confluent WM lesions
	

- Juxtaventricular WMLs (Blue)
- Periventricular WMLs (Red)
- Deep WMLs (Yellow)
- Juxtacortical WMLs (Green)



Fazekas visual rating scale for WMH
= White Matter Hyperintensity (0-6 points)

	periventricular	deep subcortical
0	absence	no or a single punctate lesion
1	„caps" or pencil-thin lining	multiple punctate lesions
2	smooth „halo"	beginning confluency of lesions
3	irregular PVH extending into deep WM	large confluent lesions

I- Multiple Sclerosis

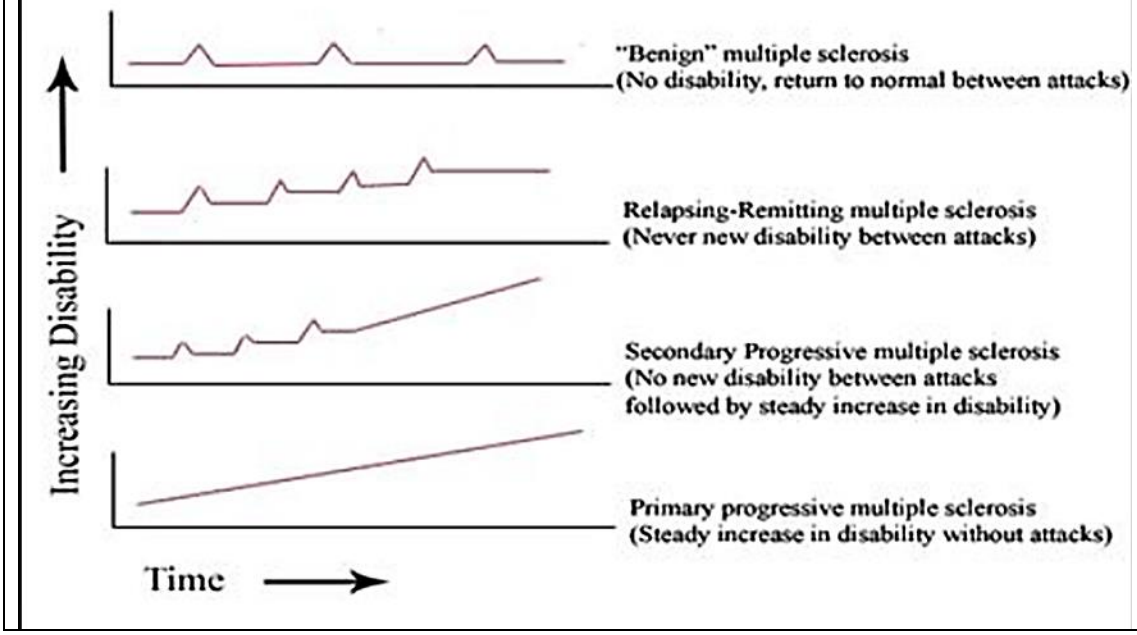
- MS is idiopathic inflammatory destruction of CNS axons in the brain and spinal cord.
- MS is one of the most common chronic demyelinating diseases.
- Young and middle aged women
- Unknown aetiology ?? Autoimmune ?? Viral mechanisms
- Familial predisposition

2 main clinical presentations of MS:

1) Relapsing–remitting (RR) (most common): Partial or complete resolution of each acute attack.

2) Progressive: No resolution or incomplete resolution between acute attacks.

- Primary progressive: Slow onset without discrete exacerbations.
- Secondary progressive: Similar to relapsing–remitting but with less complete resolution between attacks, leading to progressive disability.


"Benign" multiple sclerosis
(No disability, return to normal between attacks)
Relapsing-Remitting multiple sclerosis
(Never new disability between attacks)
Secondary Progressive multiple sclerosis
(No new disability between attacks
followed by steady increase in disability)
Primary progressive multiple sclerosis
(Steady increase in disability without attacks)
Increasing Disability
Time

❖ Mc Donald criteria for MS

The McDonald criteria for MS were recommended in 2001 by an international panel and revised in 2005.
The diagnosis is either:

- MS : all criteria fulfilled
- possible MS : not all criteria fulfilled
- not MS : no criteria fulfilled

Mc Donald criteria for MS

Dissemination in space	Dissemination in time
1 T2 lesion or more in at least two MS typical CNS regions: • Periventricular • Juxtacortical • Infratentorial • Spinal cord	• Simultaneous asymptomatic contrast-enhancing and non-enhancing lesions at any time **OR** • A new T2 and/or contrast-enhancing lesions(s) on follow-up MRI, irrespective of its timing **OR** • Await a second clinical attack

Mc Donald criteria for MS

ATTACKS	LESIONS	ADDITIONAL CRITERIA FOR DIAGNOSIS MS
2 or more	2 or more	**None.** Clinical evidence alone will suffice
2 or more	1 lesion	**Dissemination in space on MR** (or await further clinical attack implicating a different CNS site)
1 attack	2 lesions	**Dissemination in time on MR** (or await further clinical attack implicating a different CNS site)
1 attack	1 lesion	**Dissemination in space *and* time** (or await further clinical attack implicating a different CNS site)
0 attack progression from onset		One year of disease progression (retrospective or prospective) **AND** at least 2 out of 3 criteria: • Dissemination in space in the brain • Dissemination in space in the spinal cord based on 2 or more T2 lesions • Positive CSF

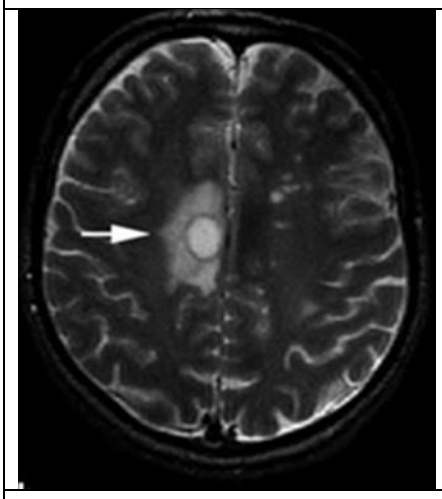
Typical Distribution:

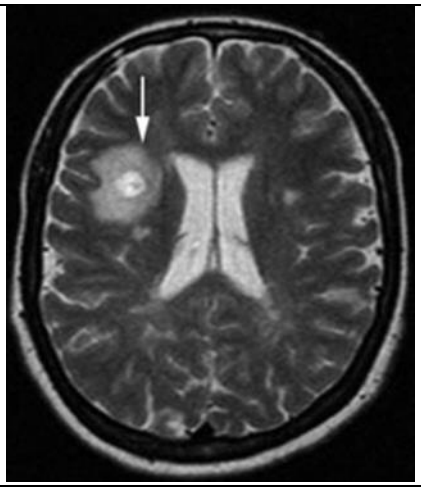
- Periventricular
- Corpus callosum
- Centrum semiovale
- Corona radita
- Internal capsule
- U fibers
- Temporal lobes
- Brain stem
- Cerebellum
- Spinal cord.

Imaging Pattern:

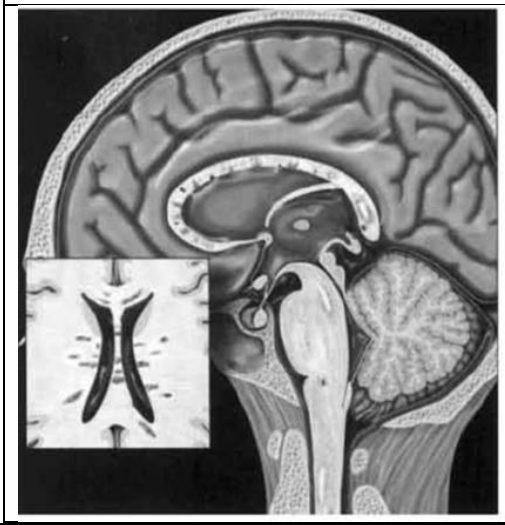
- No = Multiple
- Shape:
  - Axial images: Well marginated, rounded , oval target
  - Sagittal images: Long axis perpendicular on ventricular wall (Dawson finger)
  - Coronal images: Linear
- Size: 5-10mm up to 25mm
- T1:
  - Isointense lesions
  - Hypointense lesion = Black holes: Chronic tissue injury or severe inflammatory edema
- T2 / FLAIR: Hyperintense lesions
  - Acute lesions: surrounded by halo of edema resolve after 6m
  - Chronic lesions: No mass effect or edema
- +C:
  - Acute lesions: Enhance (Solid or ring or rim like)
  - Chronic lesions: don't enhance

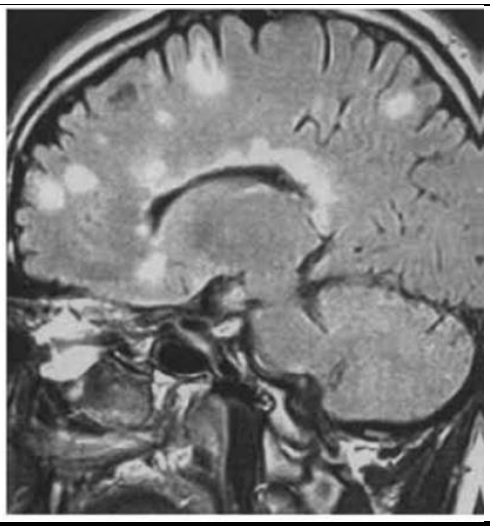
Acute MS lesions: surrounded by hallo of edema



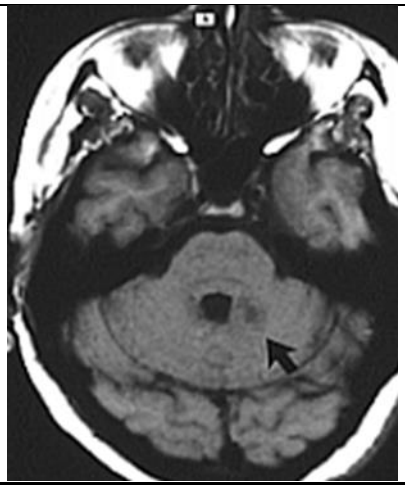


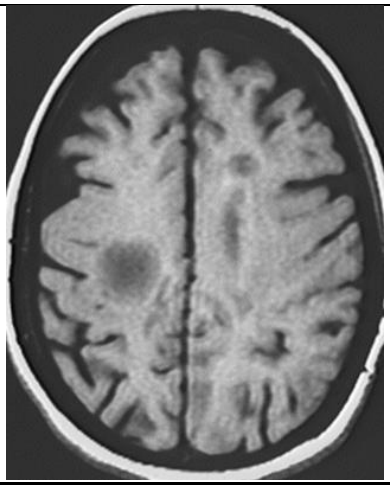
Sagittal FLAIR: hyperintense lesions with longitudinal axis perpendicular on ventricular wall => Dawson finger





T1: Black holes

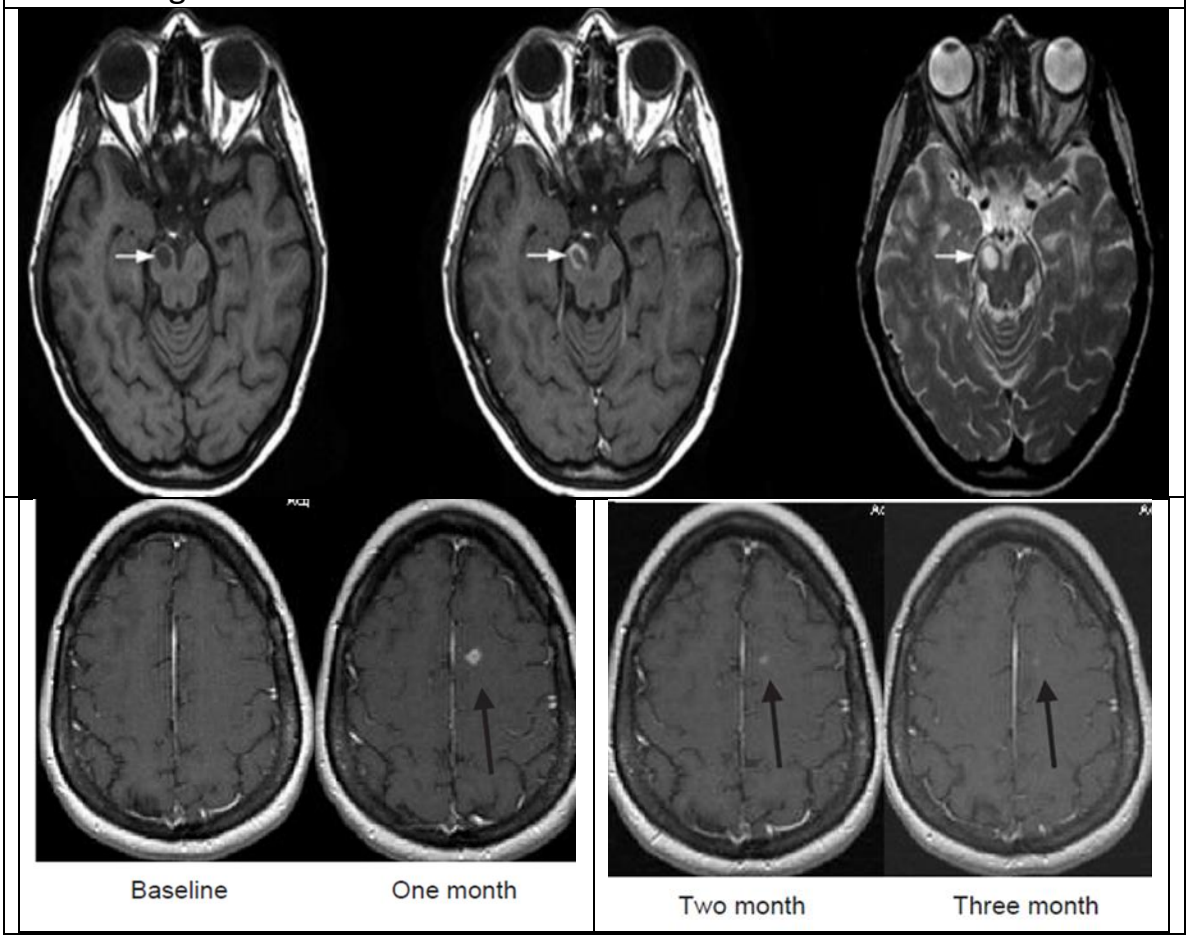




❖ Activity (inflammation) can be detected

- Post contrast T1W image
- New T2W lesions represent new inflammation
- Enlarging T2 lesions on serial studies mostly due to edema associated with inflammatory infiltrates, reaching their maximum size within 4w

T1 +C: Ring like enhancement



❖ NEW ADVANCES IN IMAGING OF MS

1) Volumetric MR Imaging: Lesion Quantification

Disease burden => based on the quantification of the T2 lesion load (ie, the total lesion volume and/or number) on MR imaging.

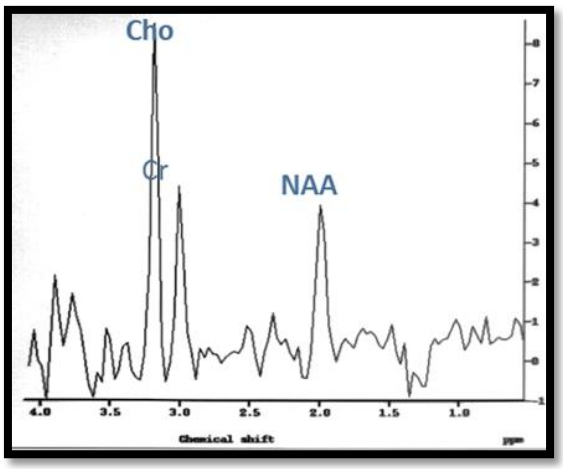
2) Magnetization Transfer

- MTI is sensitive in detecting not only the MS plaques, but also NAWM (Normal Appearing White Matter) abnormalities
- Decreased MTI have been shown in MS plaques and normally-appearing white matter (NAWM) of patients with multiple sclerosis.

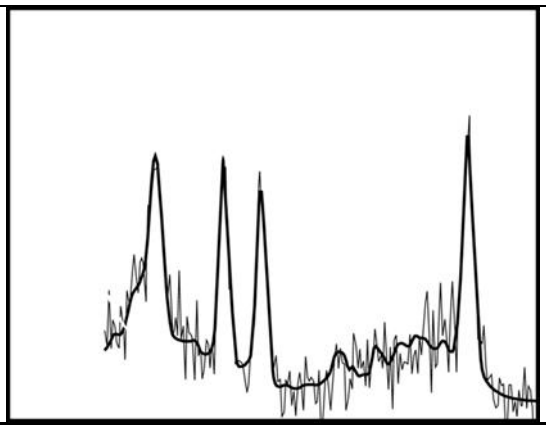
3) MRS

a) Differentiating acute from chronic plaques:

<u>Acute plaques</u>
Diminished NAA
Elevated Cho
Elevated Cho/Cr ratio
Free lipid or lactate peaks.



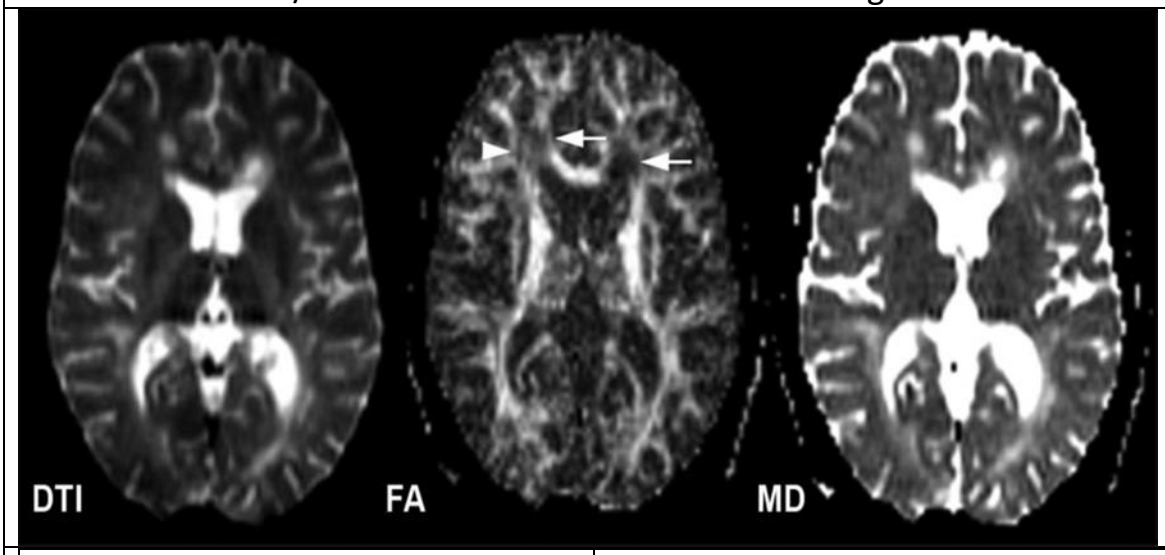
<u>Chronic plaques</u>
Diminished NAA levels
Diminished NAA/Cr ratios indicating progressive neuronal & axonal loss.
Also it may show increased mI, possibly indicating gliosis



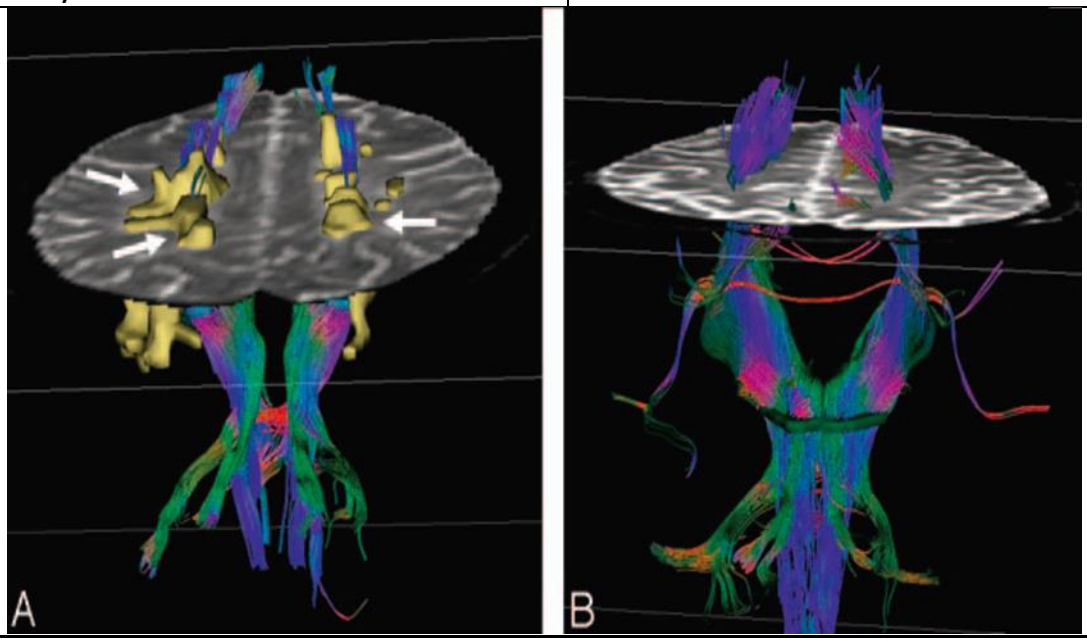
b) Differentiating RR from SP MS:
• Significant reduction of NAA/Cr and NAA/Cho ratios is observed in NAWM in patients with SP MS compared with control subjects and patients with RR MS.
c) Evaluation of occult disease:
• MRS can detect the biochemical alteration of metabolites in NAWM adjacent to or distant from lesions in MS. • The extent of NAA decrease & Cho increases in (NAWM) may progress over the time to new lesions, thus MRS provide earlier detection of MS than c-MRI. • Patients with clinically isolated syndrome (CIS) showed significant increase in NAWM myonsitol.
d) Detection of axonal damage or dysfunction & its correlation with clinical disability:
• Reduction of NAA/Cr is consistent with progressive neuronal damage. • Normalization of NAA/Cr ratio indicates neuronal recovery.
e) Monitor therapeutic response:
• Serial MRS can play a major role in the evaluation of treatment efficacy through follow up of the changes in NAA/Cr ratio & whole brain NAA level.
f) Discrimination of tumefactive demyelinating lesions from intracranial neoplasms:
• Through the significant differences in the NAA/Cr ratio in their central regions, where it is much more reduced in the neoplasms, thus avoiding unnecessary invasive procedures.
g) Discrimination between MS and age related white matter lesions:
• Elevated concentration of mI in MS plaque but not in age related white matter lesions are thought to be of special interest in their differentiation.

4) Role Of DTI in MS

- MS lesions have an increased ADC / MD and decreased FA indicates the disruption of myelin and axonal structures that leads to disorganization and increased extracellular space.
- NAWM (normal appearing white matter) have also revealed an increased ADC / MD and decreased FA in different regions



Fiber tractography in MS patient: Reduced number of fibers when they traverse white matter lesions	Fiber tractography in healthy volunteer



5) Magnetic Field Correlation (MFC), for iron quantification:

- MS patients had a significantly increased iron deposition in the basal ganglia, even in those cases with minimum signal intensity changes on conventional MR imaging

6) Perfusion MR imaging

- Decreased CBF and CBV in general
- Locally increased CBV in enhancing lesions
- Locally increased CBV in some chronic lesions

❖MS variants

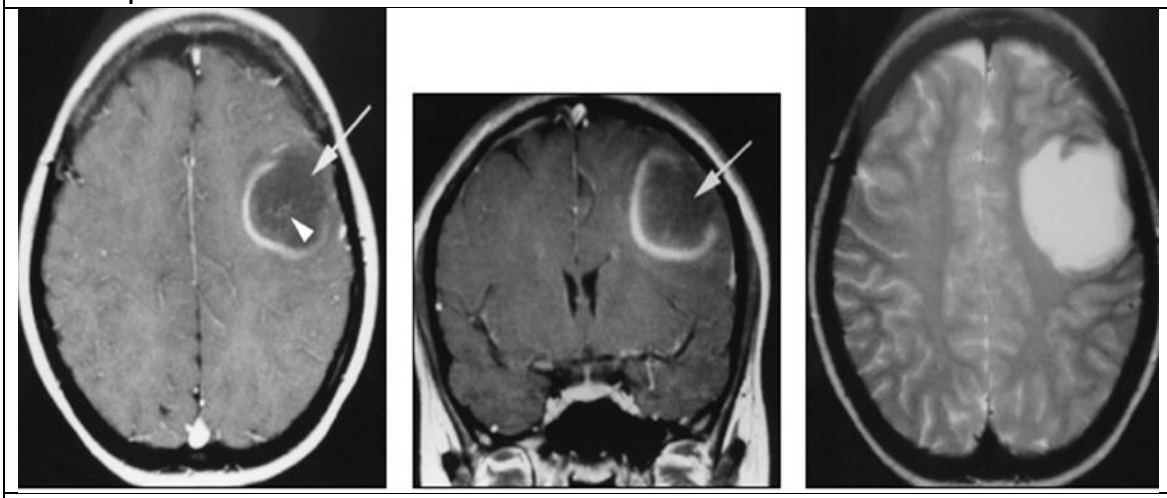
1) Tumefactive MS
2) Balo disease (1928, “encephalitis periaxialis concentrica”
3) Marburg disease (1906, acute multiple sclerosis,“encephalitis periaxialis diffusa”)
4) Schilder disease (1912, recognized as a childhood variant of Marburg encephalitis periaxialis diffusa by Schilder)
5) Devic’s disease (1894, neuromyelitis optica)

1) Tumefactive MS

- Tumefactive demyelinating lesions prove to be a diagnostic dilemma to neurosurgeons, radiologists, and pathologists.
- The clinical and imaging characteristics of these demyelinating lesions may mimic primary and secondary brain tumors, brain abscess, tuberculoma, and other inflammatory disorders e.g. sarcoidosis, primary sjogren's syndrome

Imaging findings:

- Large lesions (>2 cm.)
- White matter lesions
- Little mass-like effect or vasogenic edema
- Post-gadolinium MRI: Incomplete ring enhancement.
- MR perfusion is not increased



Corpus Callosum lesions

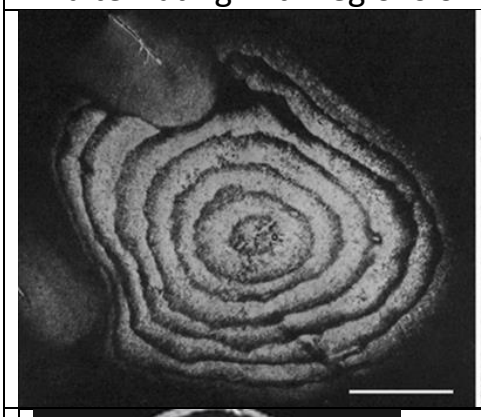
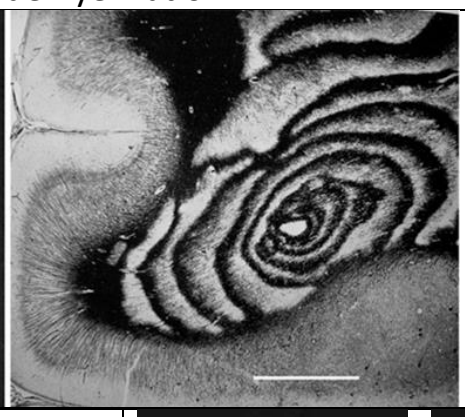
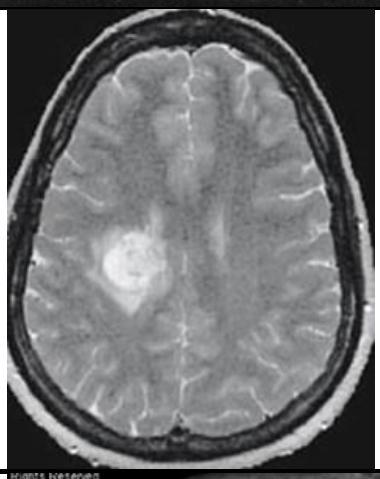
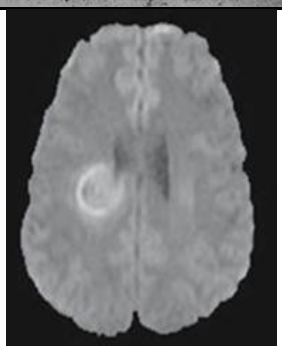
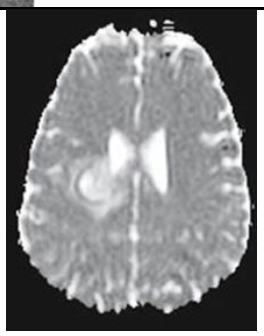
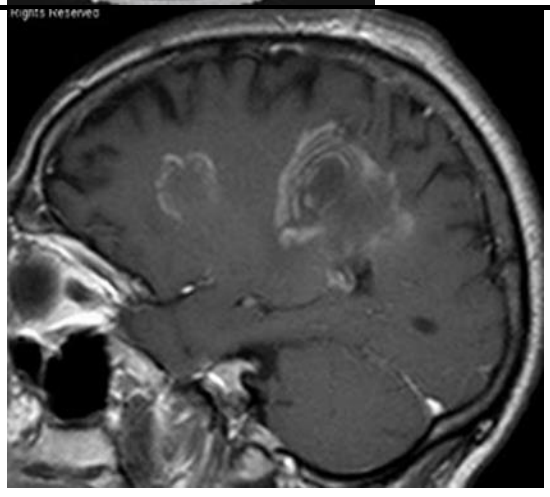
- Tumefactive demyelinating lesions can spread through or originate within The corpus callosum
- DD: "butterfly" corpus callosum lesions

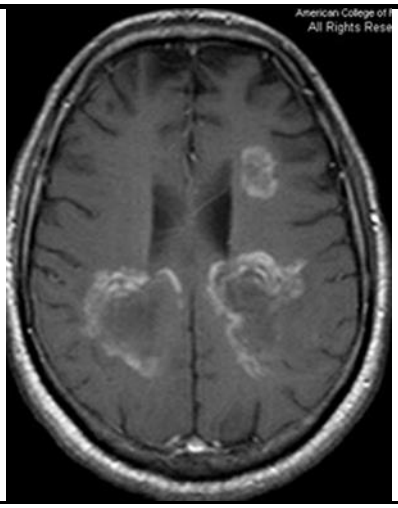
Diffusion

- Tumefactive MS: Mildly increased ADC
- Cerebral abscesses: restricted diffusion centrally within the lesion + decreased ADC
- Necrotic neoplasms may display a similar increase in ADC centrally within the lesion, making diffusion less helpful in differentiating from neoplasms

2) Balo's Concentric Sclerosis

- Peculiar pattern of pathology in cerebral hemispheric white matter consisting of a concentric, mosaic, or floral configuration of alternating bands of white matter whose basis is relatively preserved myelination alternating with regions of demyelination



3) Acute MS (Marburg Type)

- Severe, acute MS that meets both clinical and pathological criteria a monophasic illness and may best be defined by its malignant course
- Childhood
- Acute, severe, idiopathic, inflammatory demyelinating disease may be relatively unresponsive to conventional therapy (corticosteroids).
- Death may occur in weeks to months, either from severe widespread cerebral lesions or acute involvement of the lower brainstem or upper cervical cord

Imaging appearance:

- Large, often confluent lesions, more commonly affecting cerebral hemispheric white matter; sometimes involving the brainstem.
- Lesions may show enhancement and perilesional edema is often present.
- If a single lesion is the initial presentation of MS, the neuroimaging characteristic may be difficult to distinguish from a neoplasm.

4) Schilder's disease, encephalitis periaxialis diffusa

- Diffuse myelinoclastic sclerosis
- Rare , Childhood
- Essentially a diagnosis of exclusion
- Its clinical and MRI appearance may overlap with inherited metabolic disorders of myelin, particularly adrenoleukodystrophy or possibly acute disseminated encephalomyelitis

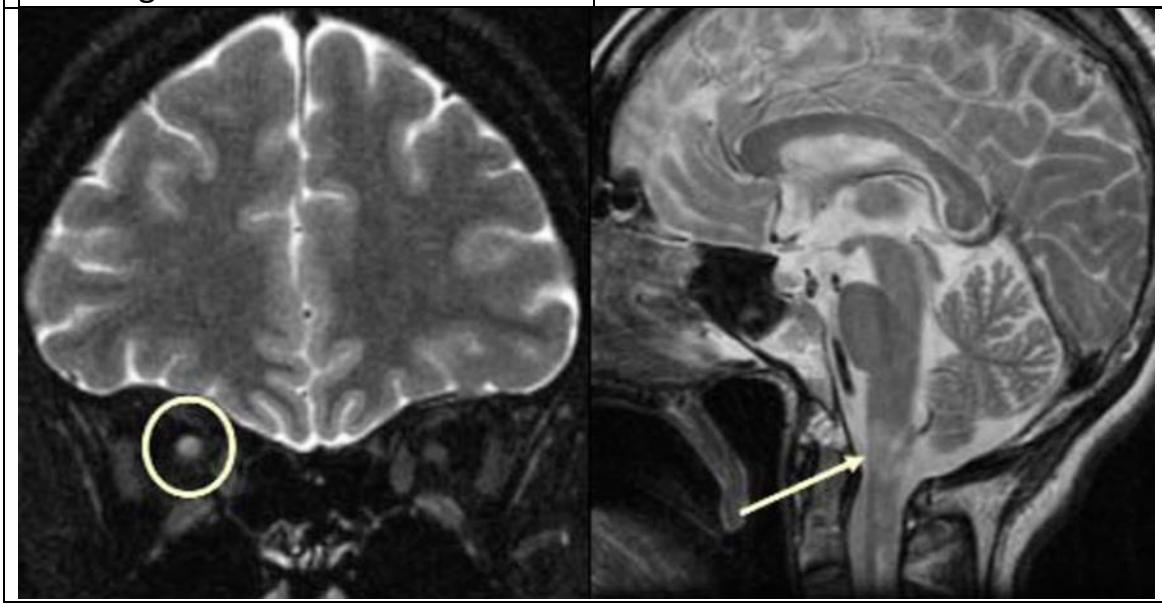
Imaging findings:

- Large, bihemispheric lesions of white matter that may show enhancement at the perimeter
- The lesions occurring in two hemispheres may be bridged by abnormal signal in the corpus callosum.
- Frank necrosis and cavitation may also occur.

5) Devic's Neuromyelitis Optica (DNO)

- The key clinical features of DNO include acute visual loss, often bilateral, and acute transverse myelitis
- Visual + spinal cord signs and symptoms often presenting nearly simultaneously
- Optic neuritis + Acute myelitis + Normal brain

Spinal cord lesions	
DNO	**MS**
Large lesions	Small lesions
Axial Section: full thickness of spinal cord (GM+WM)	Axial section: typically partial and asymmetric involvement
The vertical segment: Often exceeding three segments in height.	The vertical extent is usually two segments or less.
The spinal cord may show Striking swelling	Spinal cord swelling is relatively mild.



II- Infectious WM disease

1) Acute disseminated encephalomyelitis (ADEM)

2) Hurst's disease = Acute hemorrhagic leukoencephalitis

3) HIV-associated encephalitis

4) Progressive multifocal leukoencephalopathy (PML)

5) Subacute sclerosing panencephalitis

6) Lyme disease

1) ACUTE DISSEMINATED ENCEPHALOMYELITIS (ADEM)

- Allergic or autoimmune reaction with viral protein.
- It is usually triggered by an inflammatory response to viral infection or vaccination.
- Self limited

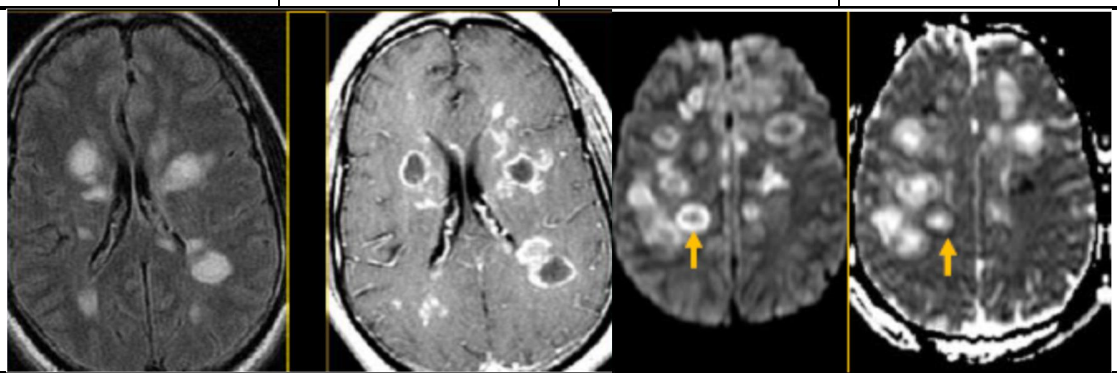
<u>Sequale:</u>

→ 80% lesions are responsive to steroids (partially or completely resolve).
→ 10-30% neurologic sequelae

<u>Imaging Findings:</u>

• Bilateral Multifocal • Large • Confluent or punctate • White matter lesions	• T1: Hypointense • T2/FLAIR: Hyperintense • T1+C: may show ring enhancement • DWI: Hyperintense • ADC: Increased

Axial T2	Axial T1+C	DWI	ADC



2) Acute Hemorrhagic Leukoencephalitis = Hurst's disease

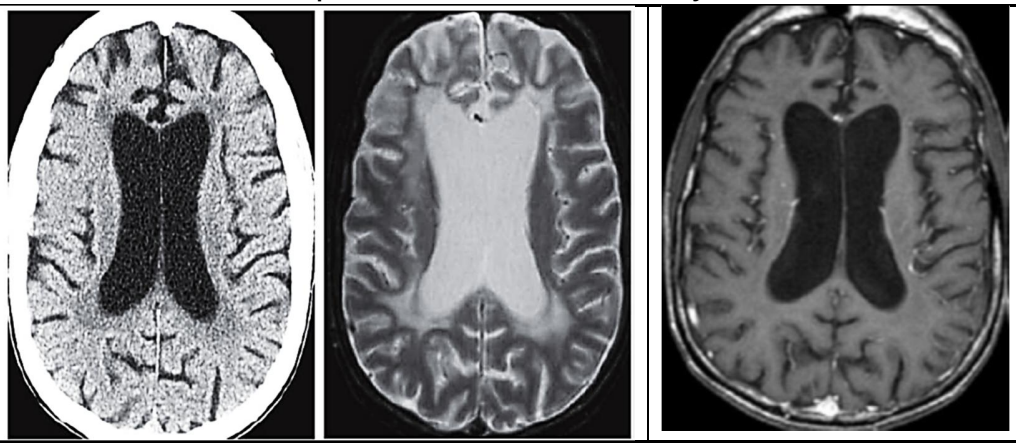
- Rapidly progressive variant of ADEM
- ADEM + Multiple WM hemorrhages
- Fatal within one week.

3) AIDS Dementia Complex
HIV Encephalopathy

- Neurodegenerative syndrome characterized by progressive
  - Cognitive and motor impairment.
  - Atrophic changes involving the brain.
  - White matter demyelination and gliosis.

<u>Imaging Findings:</u>

- Diffuse cerebral atrophy
- Bilateral symmetric
- Patchy or confluent areas
- Periventricular and deep WM.
- CT: Hypodense
- T1: Hypointense
- T2/FLAIR: Hyperintense
- T1+C: NO enhancement
- NO mass effect
- NO restricted DWI
- MRS: ↓NAA and ↑peaks of in choline and myoinositol

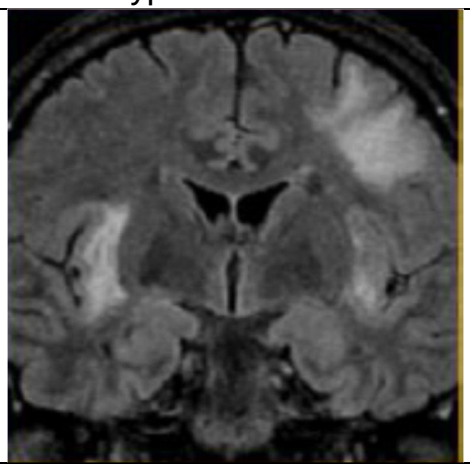
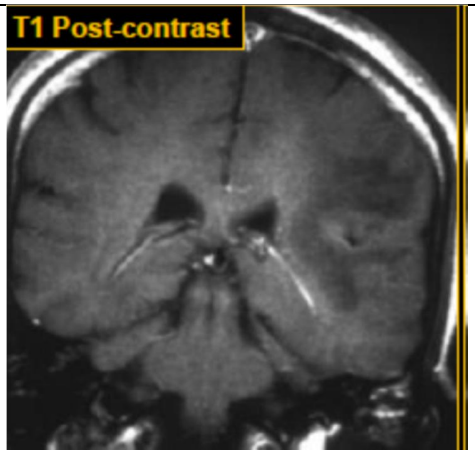


4) PROGRESSIVE MULTIFOCAL LEUKOENCEPHALOPATHY (PML)

- Fatal demyelinating disease of CNS.
- in immunocompromised patients
- AIDS patients with low CD4 count (50-100cell/mm3)
- caused by CNS infection with human JC papova viruses.

Imaging findings:

- Bilateral + Asymmetrical
- Multifocal + Patchy or Confluent
- Peri-ventricular and sub-cortical U fibers.
- CT: Hypodense
- T1: hypointense
- T2 / FLAIR: Hyperintense
- T1+C: No enhancement
- No mass effect
- DWI: Hyperintense
- ADC: Iincreased ADC
- but occasionally it shows hyperintensity with decreased ADC, especially at the margin of the lesion.

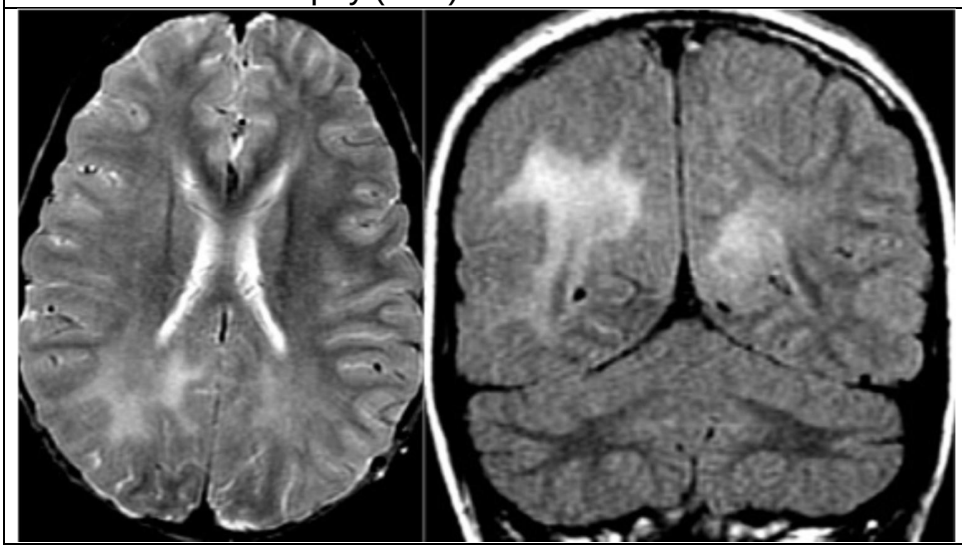
FLAIR: Hyperintense	T1+C: No enhancement
	

5) Subacute Sclerosing Panencephalitis

- Slow virus infection
- About half of patients had measles before age of 2y
- Mean age 7 years
- Insidious onset of behavior changes & mental deterioration
- Death: 1-3 years
- Diagnosis:
    - Characteristic periodic EEG discharges
    - Raised measles antibody titre in plasma & CSF

<u>Imaging findings:</u>

- May be normal 1st 3-4 months
- T2 hyperintensity
- Bilateral and asymmetric
- Parietal & occipital lobes
- Periventricular, corpus callosum, basal ganglia,
- Cerebellum & pons
- May enhance
- Generalized atrophy (Late)



III – Metabolic / Nutritional Diseases

1) **Osmotic Demyelination Syndrome**
    a. **Central pontine myelinolysis**
    b. **Extrapontine myelinolysis**
2) **Alcohol**
    a. **Marchiafava-Bignami Disease (MBD)**
    b. **Wernicke Encephalopathy**
3) **Kernicterus**
4) **B12 Deficiency**
5) **Hypothyroidism**
6) **Hypoglycemia**
7) **Nonketotic Hyperglycemia Hemichorea (NHH)**
8) **Hepatic :**
    a. **Wilson Disease**
    b. **Chronic Hepatic Degeneration / Encephalopathy**
    c. **Acute Hyperammonemia / Acute hepatic failure**

1) Osmotic Demyelination Syndrome

- Central pontine myelinolysis = demyelination of pons
- Extrapontine myelinolysis = demyelination involving extrapontine structures.
- The clinical manifestations are variable and include spastic hemiparesis, pseudobulbar palsy, decreased levels of consciousness, and coma

Patho-Physiology:

- Osmotic demyelination is associated with electrolyte imbalances (in particular, rapid overcorrection of hyponatremia) and may be seen in:
  - Chronically alcoholic patients
  - Malnourished patients.
  - Chronically debilitated organ transplant recipients
- Rapid correction of hyponatremia = Na level increases by > 12 mmol/L/d

Clinical picture:

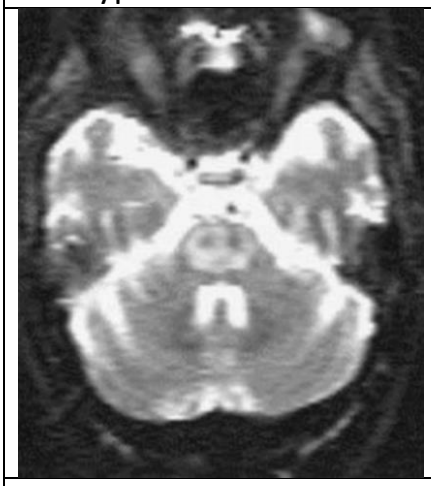
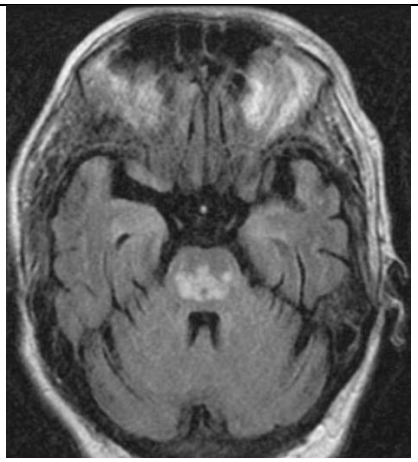
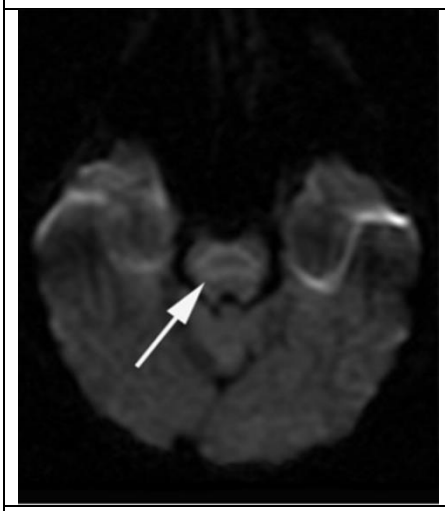
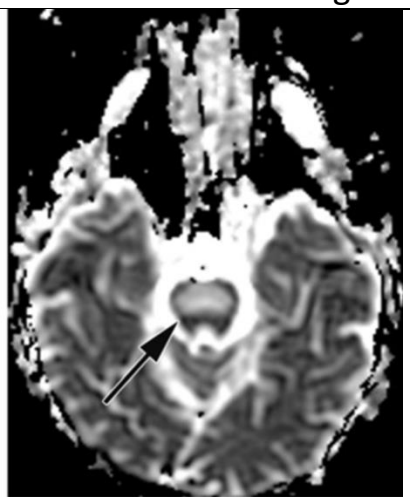
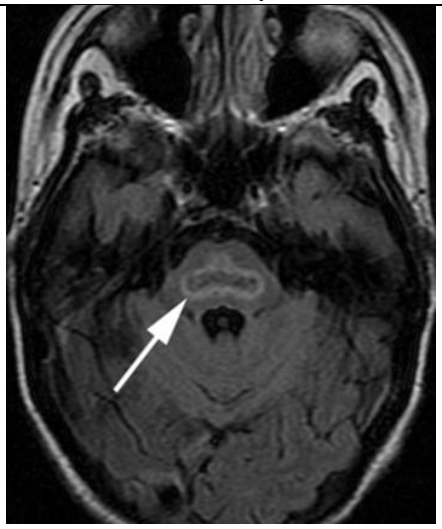
- Patients with osmotic demyelination syndrome typically present with severe electrolyte disturbances, which lead to seizures or encephalopathy.
- As normonatremia is restored, mental status improves and may return to normal within 48–72 hours.

A) Central Pontine Myelinolysis

- T1: Lesions appear isointense relative to surrounding brain tissue
- T2 & FLAIR: Symmetric **trident shaped** or **bat wing–shaped** area of increased SI in the central pons.
- +C: ± Enhance.
- Sparring ventrolateral pons and corticospinal tracts.

B) Extrapontine Myelinolysis:

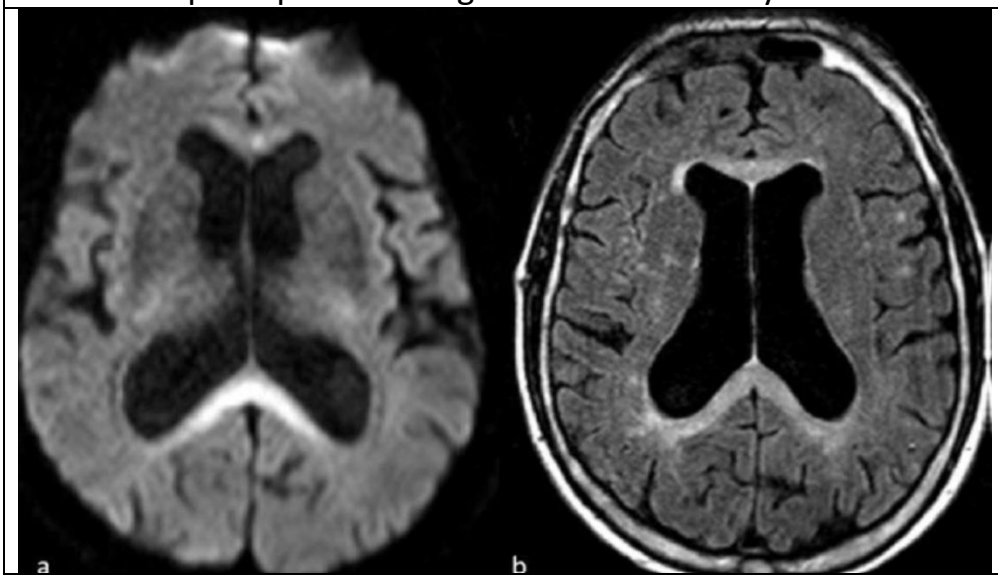
- Extrapontine myelinolysis manifests as areas of increased SI on T2 in:
  - Globus pallidus
  - Putamen
  - Thalamus
  - Cerebellum

T2: Hyperintense trident shaped	
	
DWI: Peripheral hyperintense signal	ADC: Peripheral hyperintense signal Due to T2 shine-through.
	
FLAIR: Bilateral basal ganglia + lateral thalami + Central pons	
	

2) Alcoholism

A- Marchiafava-Bignami Disease (MBD)

- is a rare CNS disorder usually seen in the context of alcoholism and malnutrition.
  - ⇨ Corpus callosum necrosis and demyelination.
- Acute phase => Corpus callosum oedematous
- Chronic phase => corpus callosum atrophy.
- Normal periaqueductal region and mammillary bodies



B- Wernicke Encephalopathy

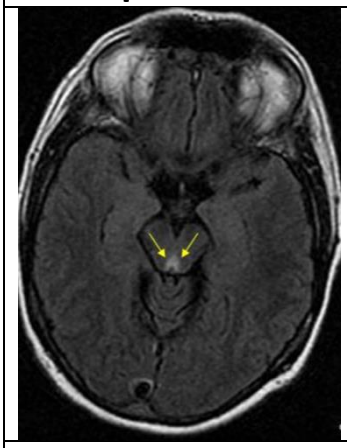
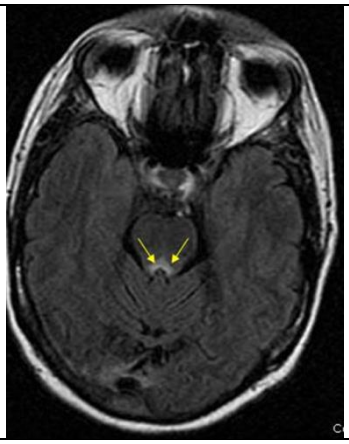
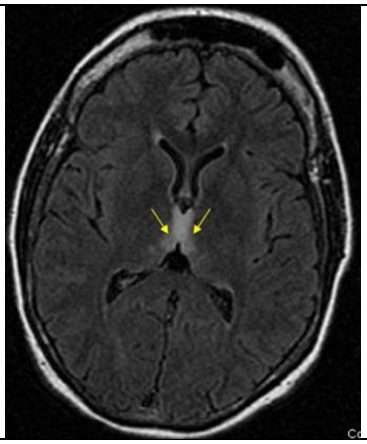
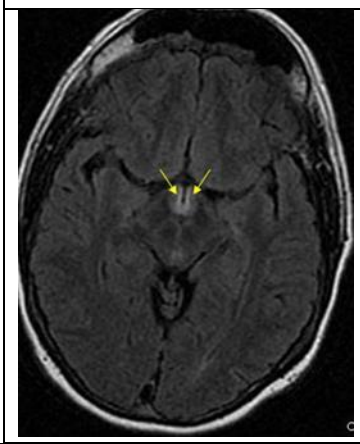
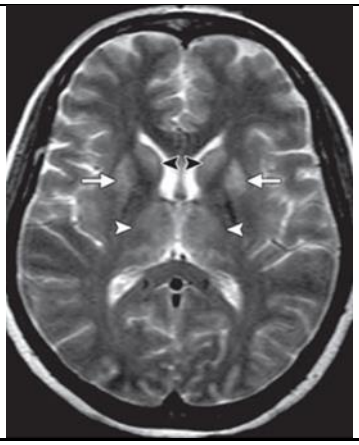
- Results from a Vitamin B1 (thiamine) deficiency, secondary to Malnourished state caused by:
  - Chronic alcoholism
  - Gastrointestinal or hematologic neoplasms
  - Chronic dialysis
  - Bowel obstruction
  - Hyperemesis gravidarum
  - Prolonged parenteral therapy without vitamin supplementation.
- Wernicke encephalopathy represents a medical emergency,
- TTT: treatment consists of intravenous replacement of thiamine

Classic Clinical Triad:

- Altered consciousness
- Ocular dysfunction
- Ataxia

Imaging findings:

- T2: Symmetric hyperintense SI in:
  - Medial thalamus
  - Periaqueductal area
  - Mamillary bodies
  - Tectal plate
- Petechial hemorrhage, diffusion restriction, and contrast enhancement of the affected areas may be noted.

Periaqueductal GM	Tectal Plate	Medial thalamus
		
Mammillary bodies	**caudate nuclei and putamina**	
		

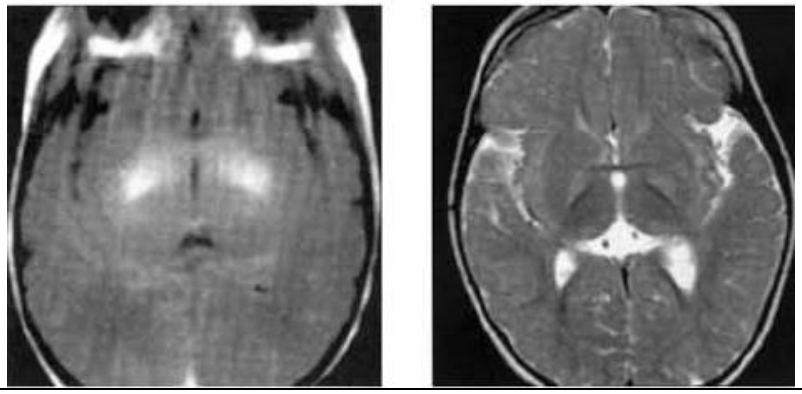
3) Kernicterus

- Encephalopathy due to deposition of toxic unconjugated bilirubin.
- ↑ Unconjugated bilirubin + immature blood-brain barrier
- Bilirubin > 20 mg/100 dLis neurotoxic in full term

- Most common cause: Erythroblastosis fetalis

Imaging findings:

- T1: High signal in GP, hippocampi, SN, DN
- T2WI: High T2 signal/volume loss in posteromedial border GP, hippocampi; occasionally DN



4) B12 Deficiency

- Most common cause in US: Pernicious anemia
- Subacute combined degeneration
- Loss of position & vibration,
- Paresthesias of hands & feet
- Lower extremity weakness
- Demyelination & axonal loss posterior & lateral spinal cord columns

Imaging Finding:

T2 / FLAIR: Hyperintense signal of posterior column of the spinal cord.

5) HYPOTHYROIDISM

- Hashimoto thyroiditis (HT)- associated ataxia
- Hashimoto encephalopathy (HE)

Imaging Findings:

Homogeneous diffuse enlargement of pituitary gland:

- ± Suprasellar extension
- ± Partial or complete obliteration of the infundibulum
- ± Compression of optic chiasma
- Enlarged pituitary gland is isointense to cerebral white matter

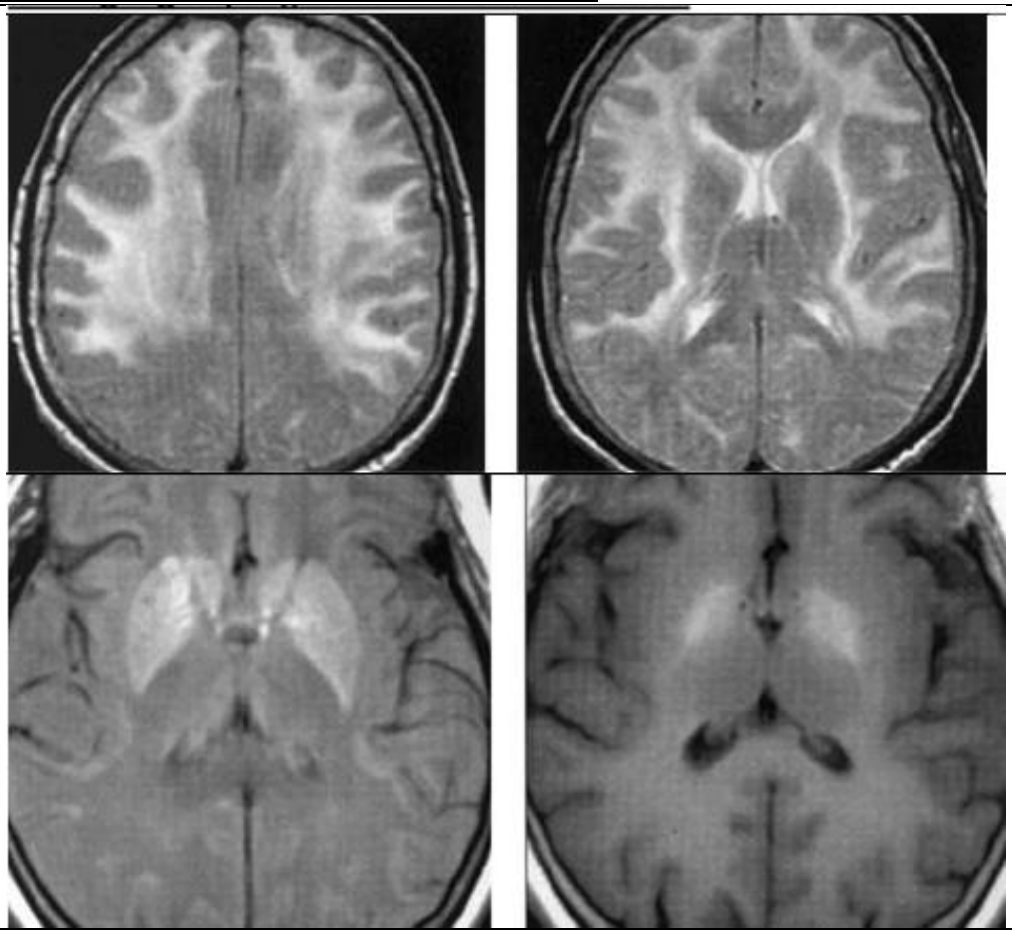
Hashimoto thyroiditis (HT)-associated ataxia

- Cerebellar vermis or olivopontocerebellar atrophy

Hashimoto Encephalopathy (HE)

- Cerebral atrophy
- Diffuse/focal cortical, subcortical WM hyperintensity

Basal ganglia (BG), variable cerebellar Ca++



6) Hypoglycemia

- Patients who present with hypoglycemic coma are typically diabetic patients who accidentally overdose while receiving treatment with oral hypoglycemic agents (typically the long-acting sulfonylurea group of drugs).
- In patients with unexplained coma, determination of blood serum sugar levels can help differentiate this potentially reversible condition from other causes such as hypoxic ischemic encephalopathy (HIE) or acute cerebral infarction

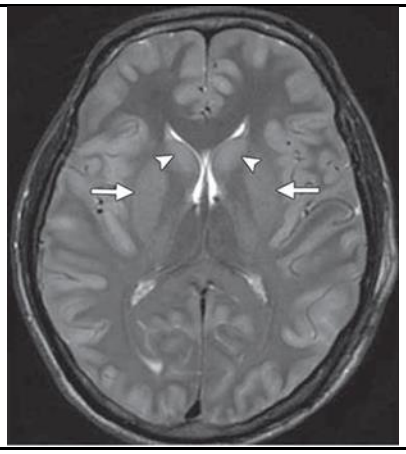
MR imaging findings:

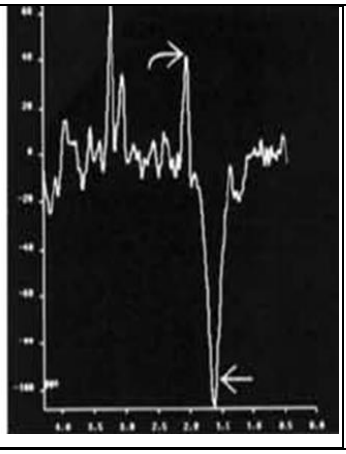
In severe cases:

- T2: bilateral hyperintense T2 signal in the cerebral cortex, hippocampi, and basal ganglia.
- Severe occipito-parietal edema or infarctions in a newborn with seizures

In mild cases:

- Transient and isolated white matter abnormalities involving the splenium of the corpus callosum, internal capsules, and corona radiata.
- DWI: The abnormal areas are typically hyperintense.
- MRS: ↓ NAA, ↑ lactate





7) Nonketotic Hyperglycemia Hemichorea (NHH)

- Patients with hyperglycemia are typically individuals with poorly controlled diabetes who present with acute chorea, hemiballismus, and, sometimes, altered mental status.
- Hyperglycemia is a treatable condition with a good prognosis
- Follow-up neuroimaging usually shows the resolution of findings.

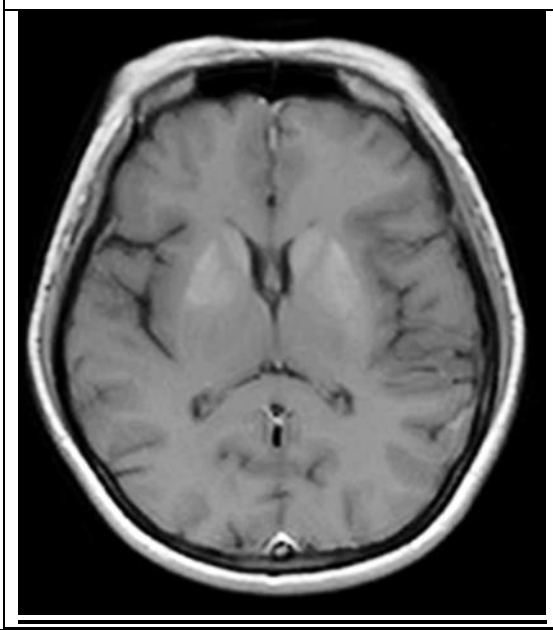
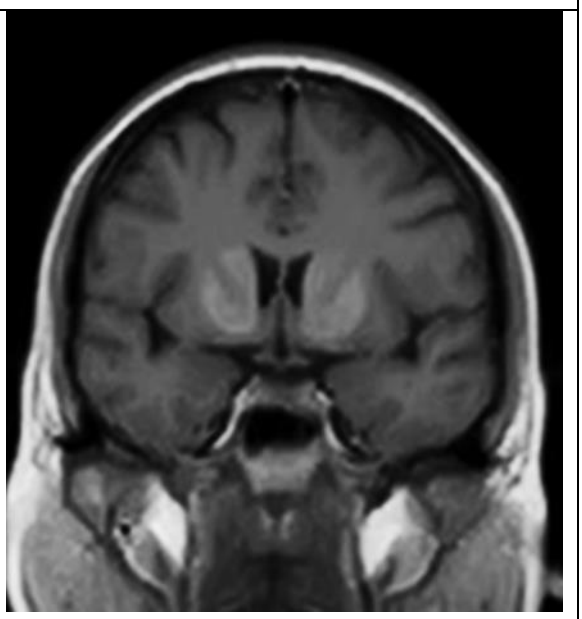
Imaging findings:

T1: bilateral hyperintense globus pallidus

T2: variable intensity.

T2/FLAIR: hypointense

DWI: shows restricted diffusion

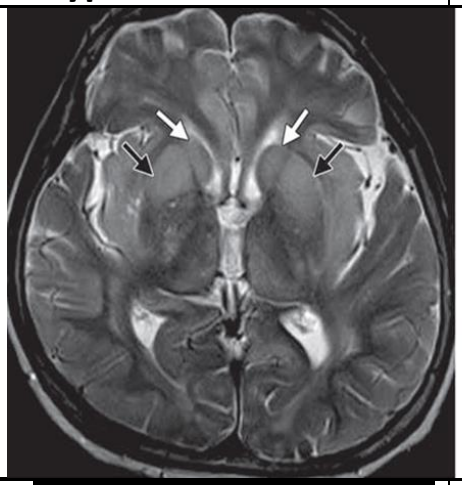
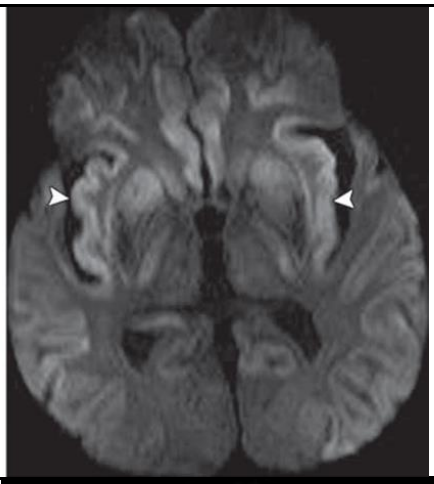
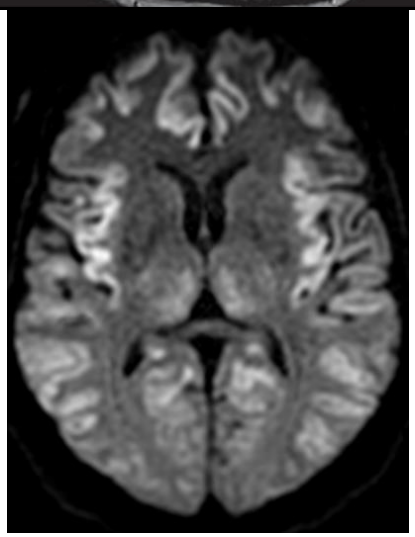
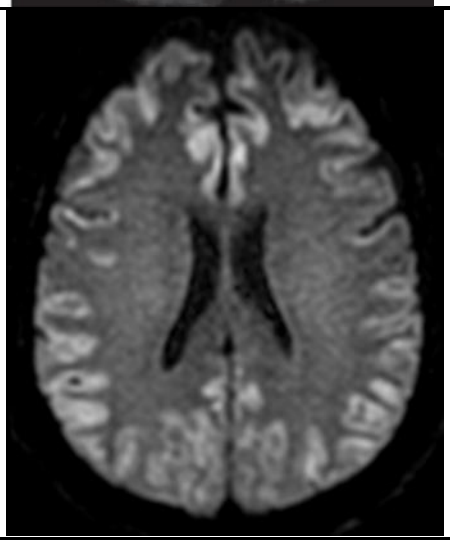
Axial T1	Coronal T1
	

8) Acute Hyperammonemia
Acute Hepatic Encephalopathy

A) Cirrhotic patients with acute hepatic decompensation (in whom the ammonia concentration can suddenly increase fourfold)
B) Patients with ornithine transcarbamylase deficiency (ie, inborn errors of metabolism such as citrullinemia, which result in accumulation of ammonia in the brain)

<u>High T2 / FLAIR signal + Restricted DWI:</u>

- Cerebral cortex, sparing perirolandic and occipital regions
- Insular cortex, Cingulate gyrus.
- Periventricular white matter (WM)
- Basal ganglia and Dentate nucleus

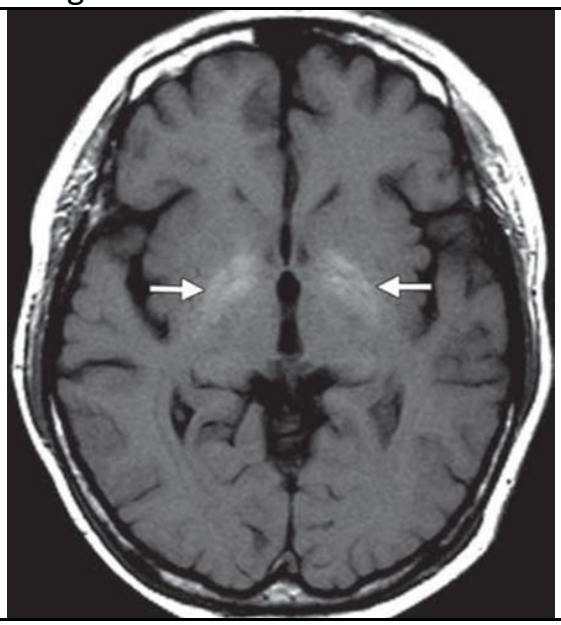
Axial T2 Hyperintense	**Axial DWI Restricted**
	
	

9) Chronic Hepatic Encephalopathy Acquired Non-Wilsonian Hepatocerebral Degeneration

- Most patients have a history of chronic cirrhosis with portal hypertension and iatrogenic (ie, placed with transjugular intrahepatic portosystemic shunt surgery) or spontaneous portosystemic shunts, resulting in nitrogenous waste products crossing the blood-brain barrier and causing long-term toxic brain damage.

Imaging findings:

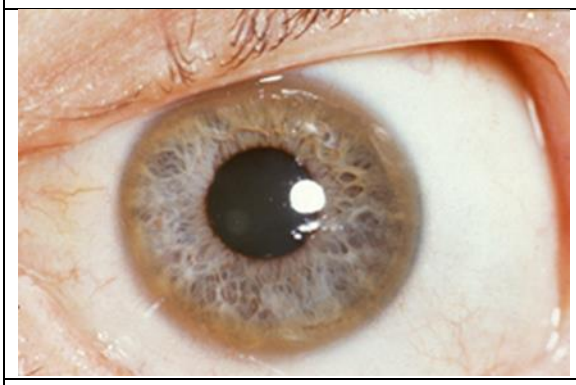
T1: Bilateral hyperintense globus pallidus and substantia nigra.



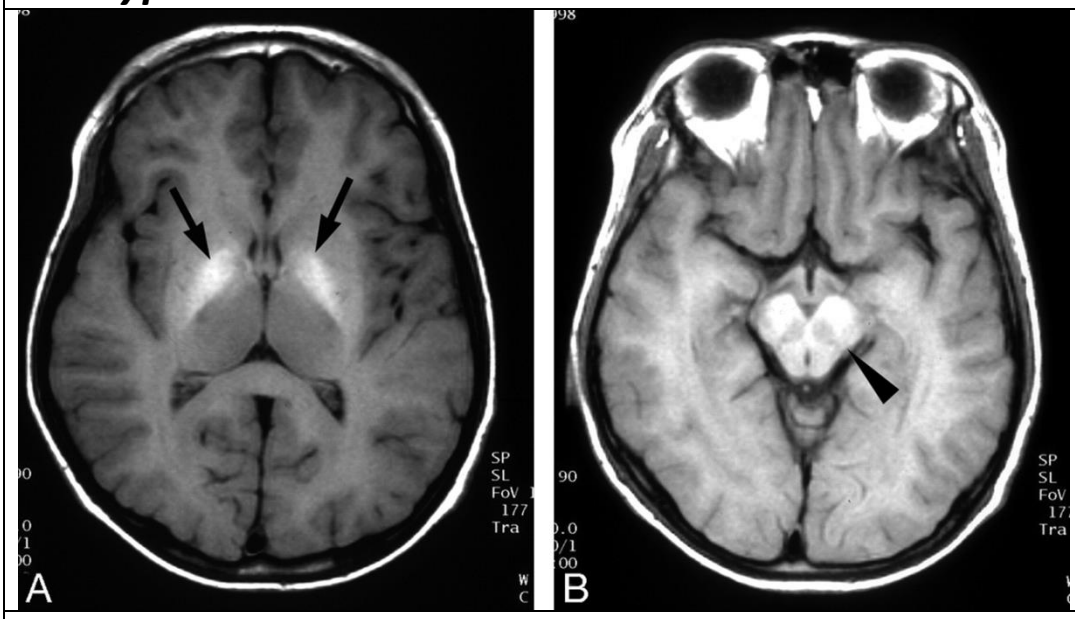
10) Wilson Disease Hepatolenticular Degeneration

- Uncommon autosomal recessive, inborn defect in copper metabolism
- Also known as hepatolenticular degeneration, affects the liver, brain, and other tissues.
- The symptoms vary and include dysarthria, dystonia, tremors, ataxia, Parkinsonian symptoms, and psychiatric problems.

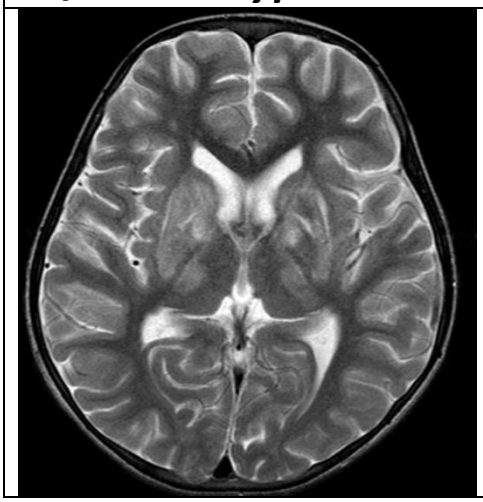
Kayser-Fleisher rings in the cornea are characteristically associated with Wilson disease.

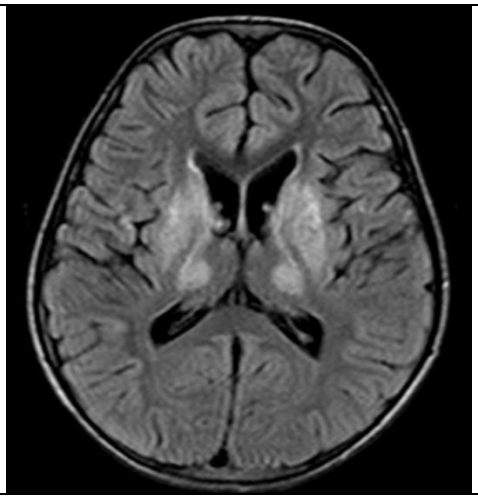


T1: Hyperintense BG + MB

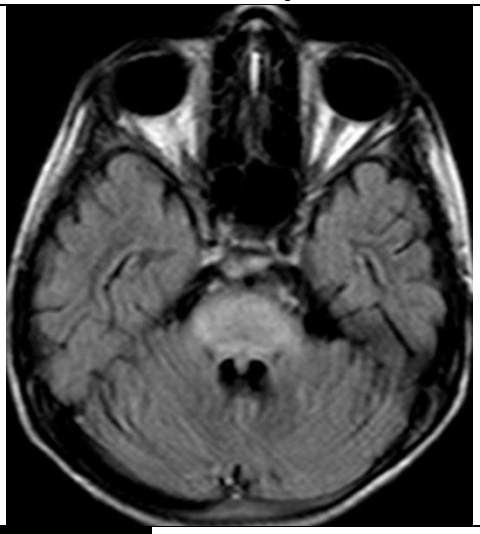


T2/FLAIR: Hyperintense BG + Anterolateral thalami



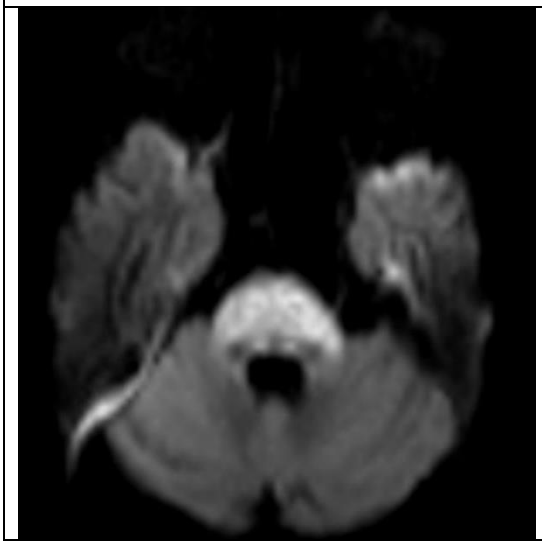


T2/FLAIR: Hyperintense Pons + Middle cerebellar peduncles.

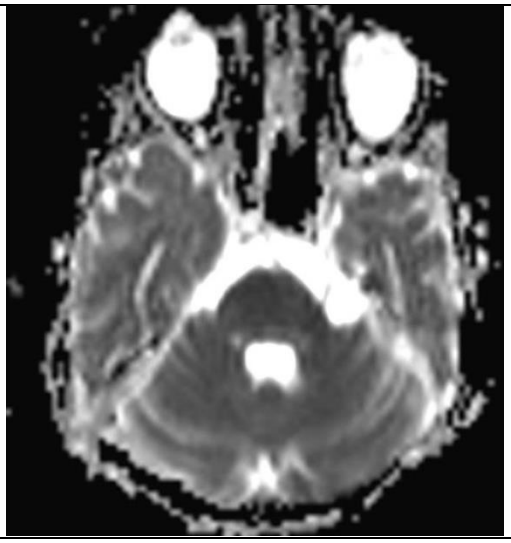


T2 MRI the "face of the giant panda" in the midbrain (arrow).

Restricted DWI



Low ADC



IV- Vascular White Matter Disease

1) Senescent white matter changes
2) Cerebral autosomal dominant arteriopathy with subcortical infarction & leukoencephalopathy (CADASIL)
3) Binswanger Disease = Subcortical Arteriosclerotic Encephalopathy (SAE)
4) Posterior reversible encephalopathy syndrome (PRES)
5) CNS Vasculitis

1) Senescent WM Changes

- White matter abnormalities
- 30-80% normal elderly
- Do not involve corpus callosum
- "Capping" of lateral ventricles
- Patchy T2 signal
- Linear T2 along body lateral ventricles
- Binswanger's: Reserved for dementia cases

2) Binswanger Disease = CHRONIC HYPERTENSIVE ENCEPHALOPATHY Subcortical Arteriosclerotic Encephalopathy (SAE)

- Is form of vascular dementia.
- Severe arteriosclerosis of the small penetrating arteries, which are thickened, hyalinized, stenotic, or even occluded.

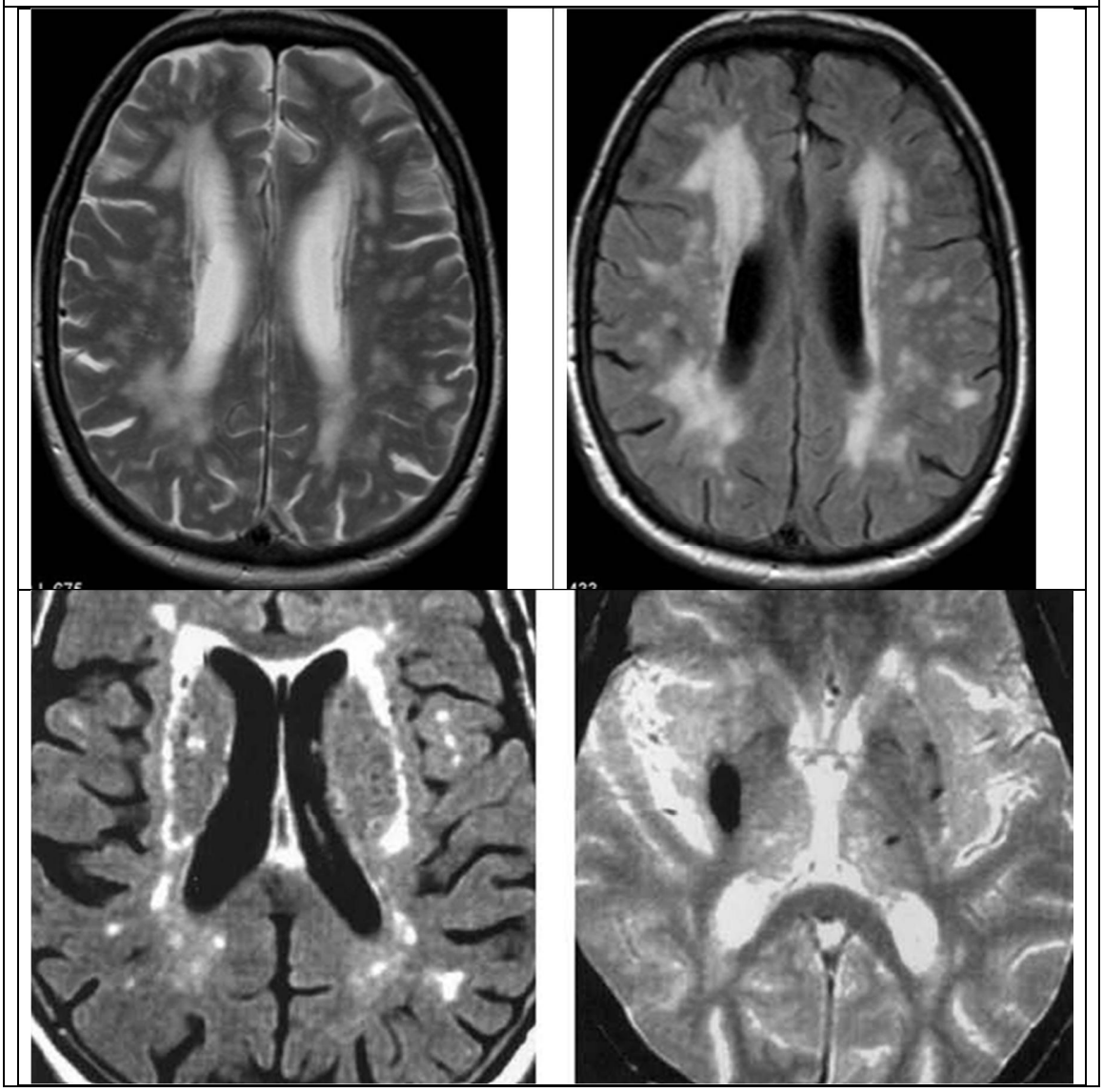
<u>Clinical Picture:</u>

- Binswanger's disease is a clinicopathologic diagnosis.
- Hypertension
- Dementia
- Pseudobulbar state
- Gait disorder often of Parkinsonian type,
- TIA = transient ischemic attack
- Multiple strokes

<u>Imaging findings:</u>

- Diffuse, confluent, periventricular hyperintense T2 / FLAIR sheets of the central white matter, especially prominent around the frontal and occipital horns and in the centrum semiovale.
- Lacunar infarcts are common in:
    - Deep central gray matter structures (BG and thalamus)
    - Corona radiata and centrum semiovale.

- Moderate cerebral atrophy.
- Petechial hemorrhage:
    - Hyperintense in T1
    - hypointense in T2*
    - Predilection for BG and thalami



3) CADASIL

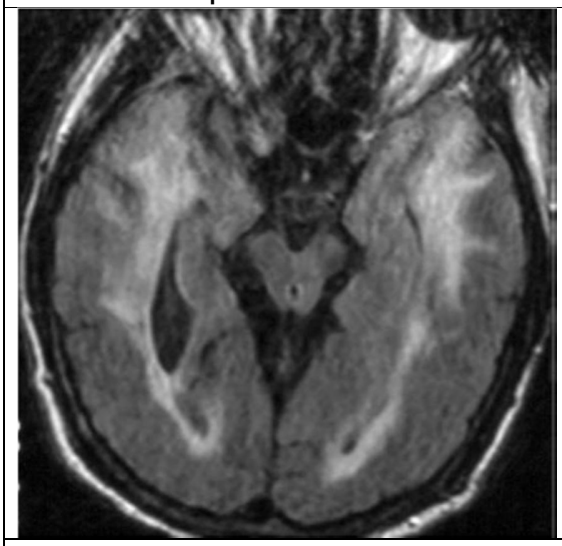
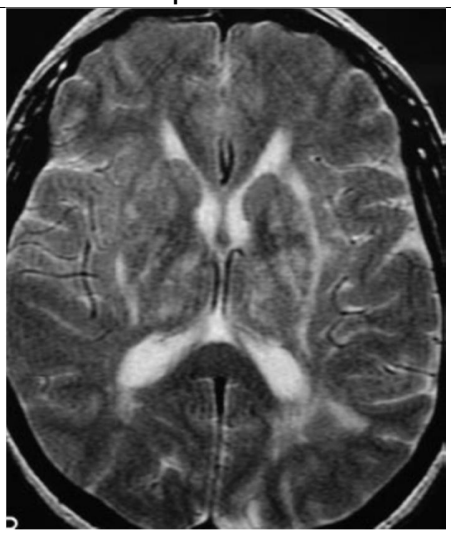
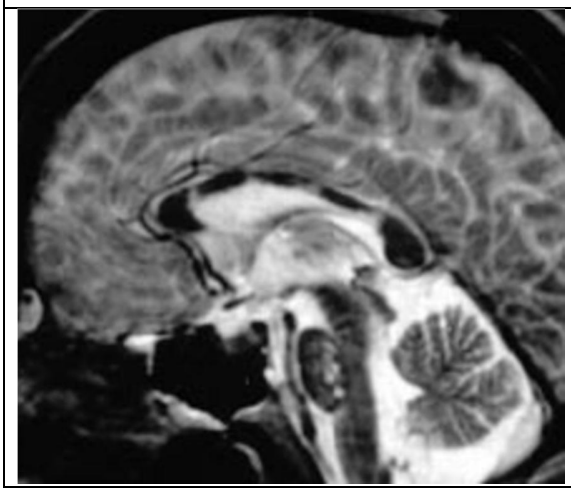
- Cerebral autosomal dominant arteriopathy with subcortical infarcts and Leucoencephalopathy
- Most common heritable cause of stroke and vascular dementia in adults.
- Mutation of Notch 3 gene on chromosome 19
- Affects frontal lobes, temporal lobes, & insula
- Skin biopsy positive 50% of time (electron dense granules in media of arterioles)

Clinical Picture :

- Early attacks of migraine with aura (<age 30)
- Later on (>age 40): TIA, strokes, progressive subcortical dementia, and mood disturbances.
- Early onset ischemic stroke (40-50's)

Imaging findings:

- Early stage of CADASIL = similar to multiple sclerosis (MS).
- Advanced stages of CADASIL = similar to Binswanger's disease

Advanced CADASIL:	
Characteristic WM Hyperintensity in Anterior temporal lobe	Characteristic WM Hyperintensity in External Capsule
	
Characteristic WM Hyperintensity in Corpus callosum	Diffuse & Confluent periventricular WM hyperintensity + T2* Microbleeds
	

4) PRES = Posterior Reversible Encephalopathy

- A disorder of cerebral vascular autoregulation
- Symptoms: Headache, seizures, confusion, drowsiness & visual disturbance

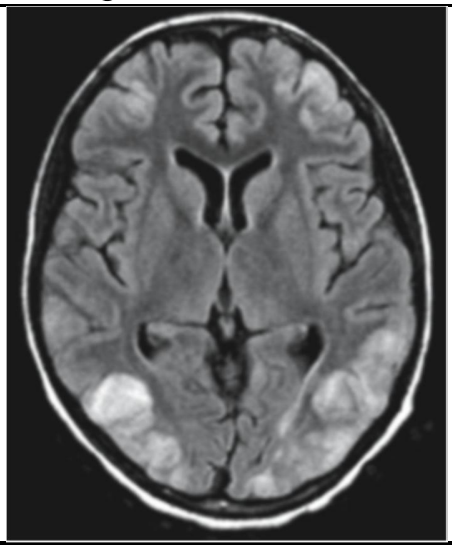
- 2 pathophysiologic mechanisms proposed:
  - Cerebral vasospasm with resulting ischemia
  - Hyperperfusion due to breakdown of cerebral autoregulation with interstitial extravasation of fluid

PRES: Associations

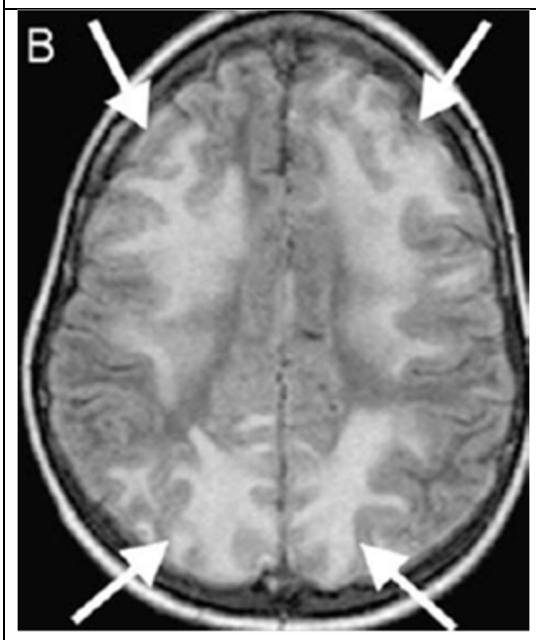
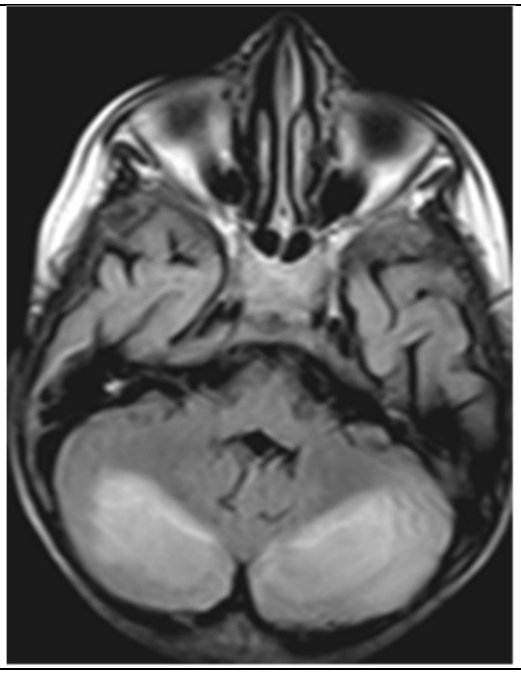
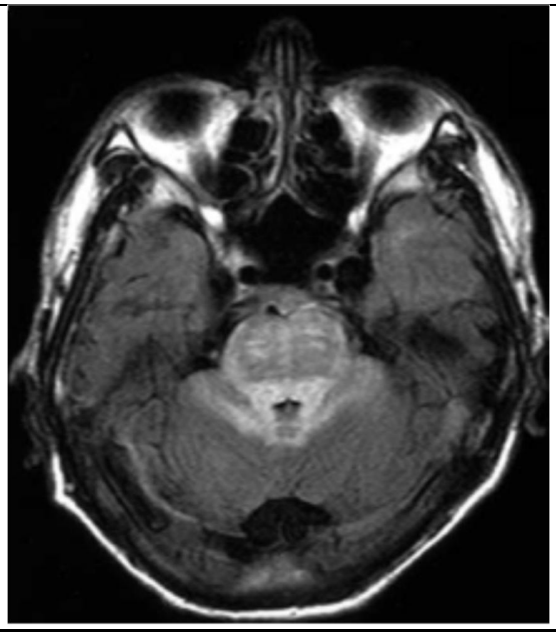
- Glomerulonephritis
- Preeclampsia & eclampsia
- SLE
- Thrombotic thrombocytopenic purpura
- Hemolytic-uremic syndrome
- Drug toxicity
  - Cyclosporine
  - Tacrolimus
  - Cisplatin
  - Erythropoietin
  - Cocaine/amphetamine

❖Typical Radiological Findings of PRES:

- Cortical and subcortical white matter lesions.
- Bilateral and asymmetric,
- Predominantly in the posterior circulation areas (parietal, occipital lobes and cerebellum)
- Hyperintense signal on T2 and FLAIR
- Isointense DWI and hyperintesne ADC => Reversible
- Resolution of MRI findings may occur as early as 1 week.



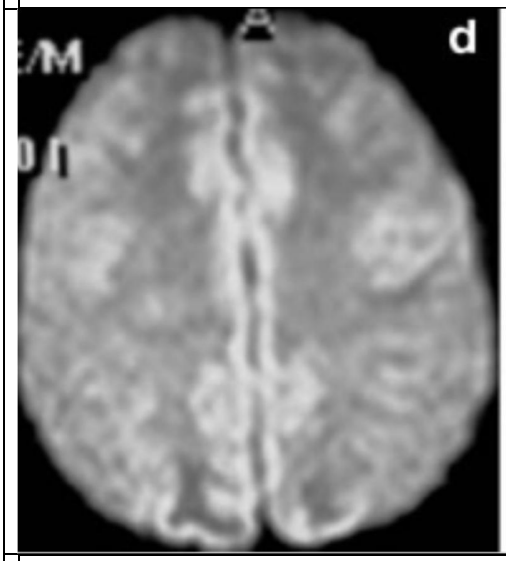
❖Atypical Locations of PRES

Extensive frontal + Parietal	Cerebellar
	
Brain Stem	**Basal ganglia**
	

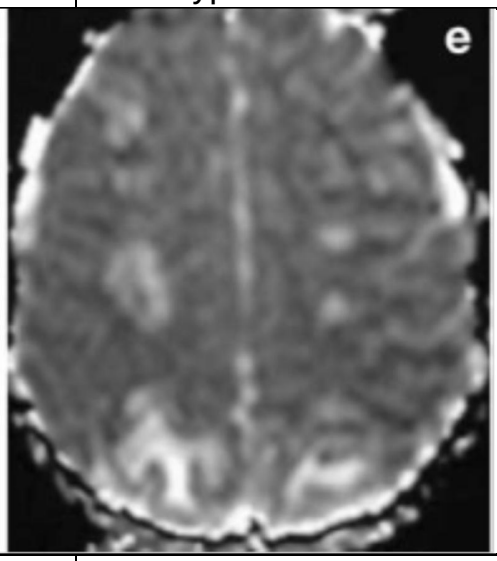
❖ Role of DWI and ADC:

	Vasogenic edema	Cytotoxic edema
DWI	↑ Increased signal intensity	↑↑ Increased signal intensity
ADC	=↑ Increased signal intensity	↓ Decreased signal intensity
Evolution	Usually reversible	Usually irreversible

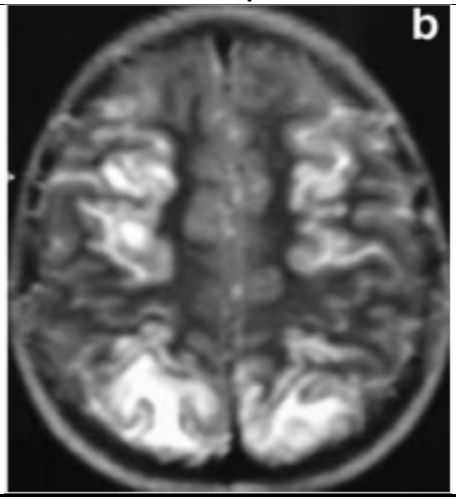
DWI: Isointense



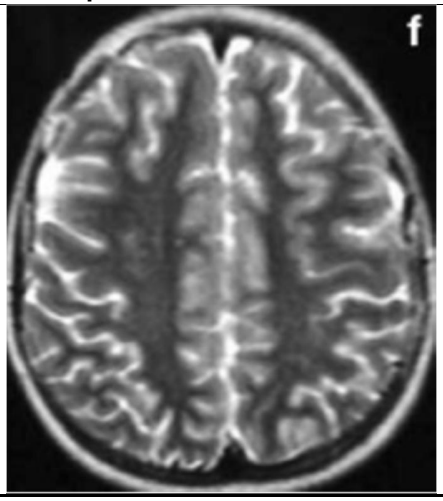
ADC: Hyperintense



❖ Follow up =>



Complete resolution



5) CNS Vasculitis

- Heterogeneous group of inflammatory disorders

Table 1: Classification of Vasculitis according to the 2012 Revised International Chapel Hill Consensus Conference on the Nomenclature of Systemic Vasculitides

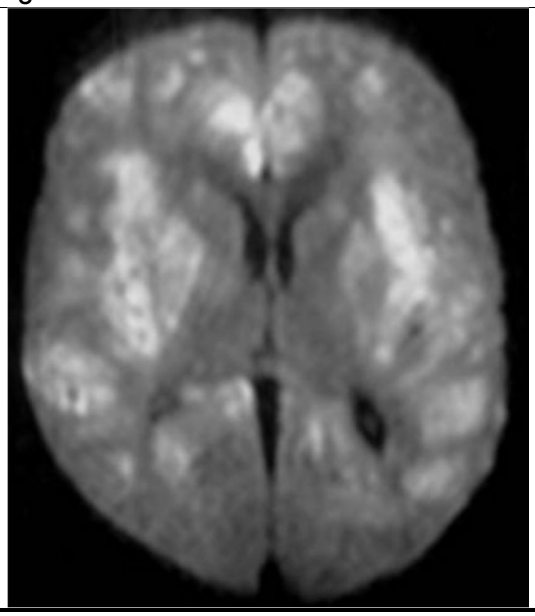
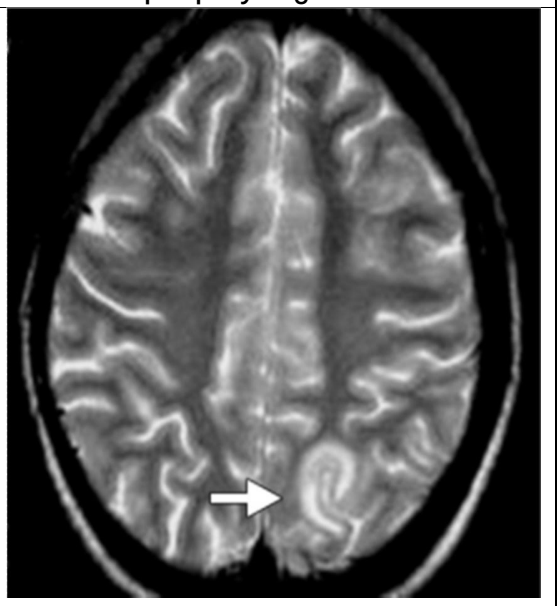
Type of Vasculitis	Disease Entities
Large-vessel vasculitis	Takayasu arteritis Giant cell arteritis
Medium-sized vessel vasculitis	Polyarteritis nodosa Kawasaki disease
Small-vessel vasculitis	IgA vasculitis Microscopic polyangiitis Granulomatosis with polyangiitis Eosinophilic granulomatosis with polyangiitis
Variable-sized vessel vasculitis	Behçet disease Cogan syndrome
Single-organ vasculitis	PACNS
Vasculitis associated with systemic disease	SLE Sjögren syndrome Rheumatoid arthritis APLA syndrome Scleroderma
Vasculitis associated with probable etiology	Infection-induced vasculitis Acute septic meningitis *Mycobacterium tuberculosis* Neurosyphilis Viral (HIV-related vasculitis, varicella-zoster vasculopathy) Fungal (mucormycosis, aspergillosis) Parasitic (cysticercosis) Malignancy-induced vasculitis Drug-induced vasculitis Radiation-induced vasculitis

<u>Direct Imaging findings = Vascular changes (Angiography / CTA / MRA):</u>

- Vessel wall thickening and contrast enhancement
- Stenosis / occlusion
- Aneurysms
- Segmental narrowing & dilatation

Indirect Imaging findings = Parenchymal changes:

- Multiple areas of T2 hyperintensity
- Deep & subcortical white matter
- Patchy or confluent
- ± Enhance
- May appear identical to MS
- Micro or macro- Infarction
- Micro or macro- hemorrhage

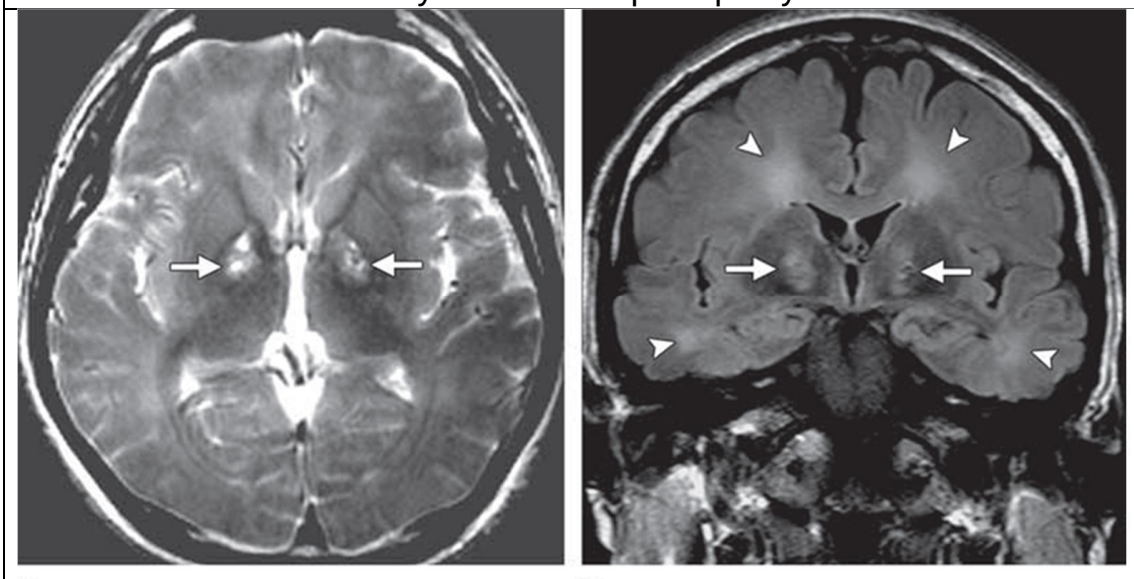
IgA vasculitis	Microscopic polyangiitis
	

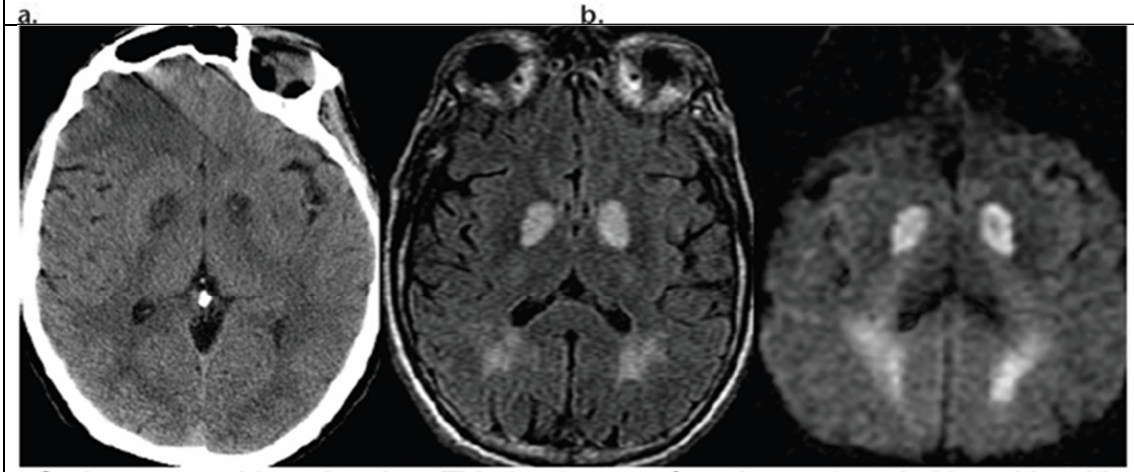
V- Toxic Poisoning

1) Carbon Monoxide Poisoning

<u>Imaging findings:</u>

- T2 & FLAIR: symmetric hyperintense foci in globus pallidus (arrows).
- Symmetric hyperintense areas in deep white matter (arrowheads in b) are consistent with delayed leukoencephalopathy.

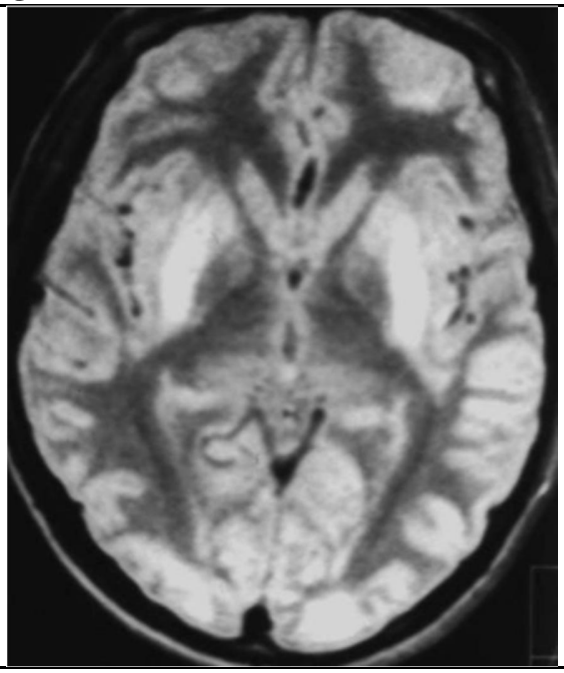




Carbon monoxide poisoning. This hunter was found comatose in his cabin with a propane heater still running. There is focal necrosis of the globi pallidi and early white matter demyelination. Case courtesy of Dr. Dick Latchaw

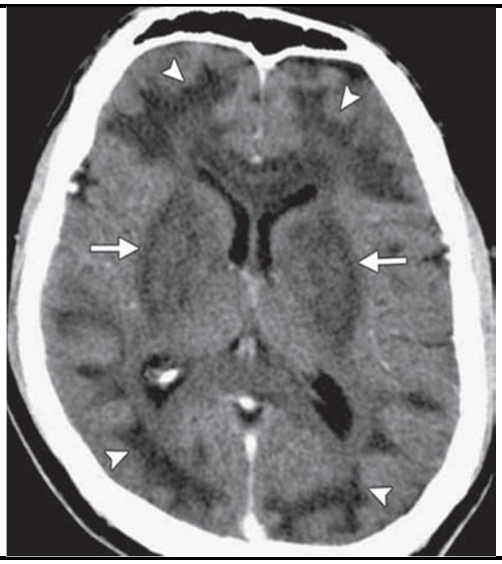
2) Methanol Poisoning

Bilateral T2 hyperintensity in the putamen



3) Toxic Encephalopathy

- CT hypodense
- T2 / FLAIR: Hyperintense
- Patchy / confluent areas at:
- Lentiform nuclei (arrows)
- Corpus callosum,
- Subcortical deep white matter in the frontal and parietooccipital regions (arrowheads).

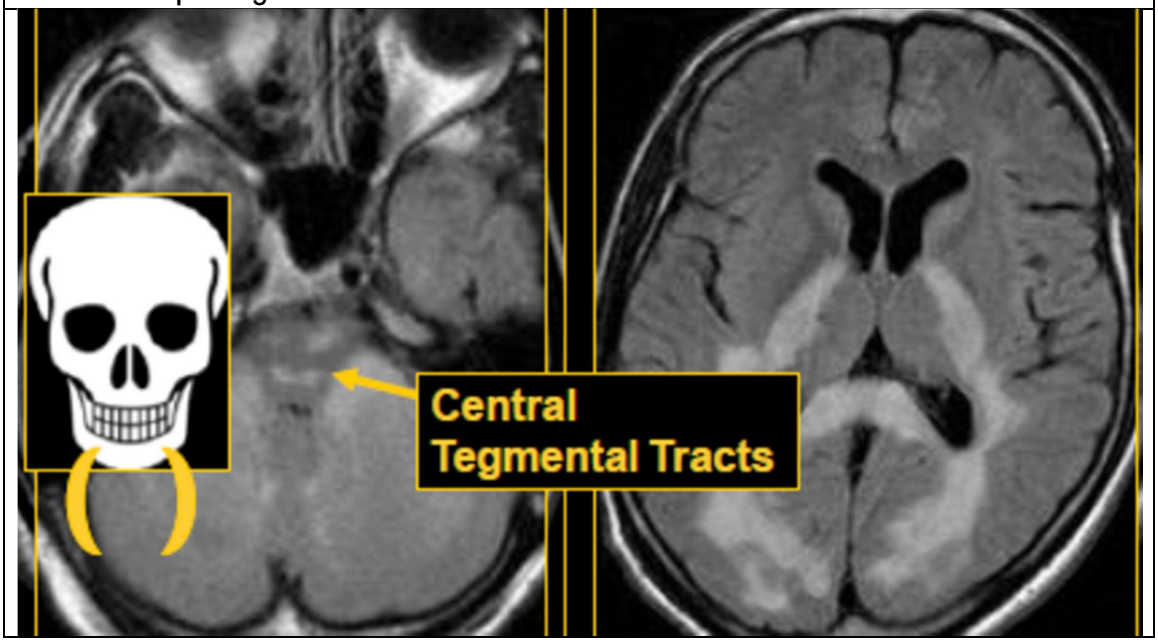


VI- DRUG ABUSE

- Young/middle-aged adult with ischemic or hemorrhagic stroke occurring in close temporal proximity to drug administration

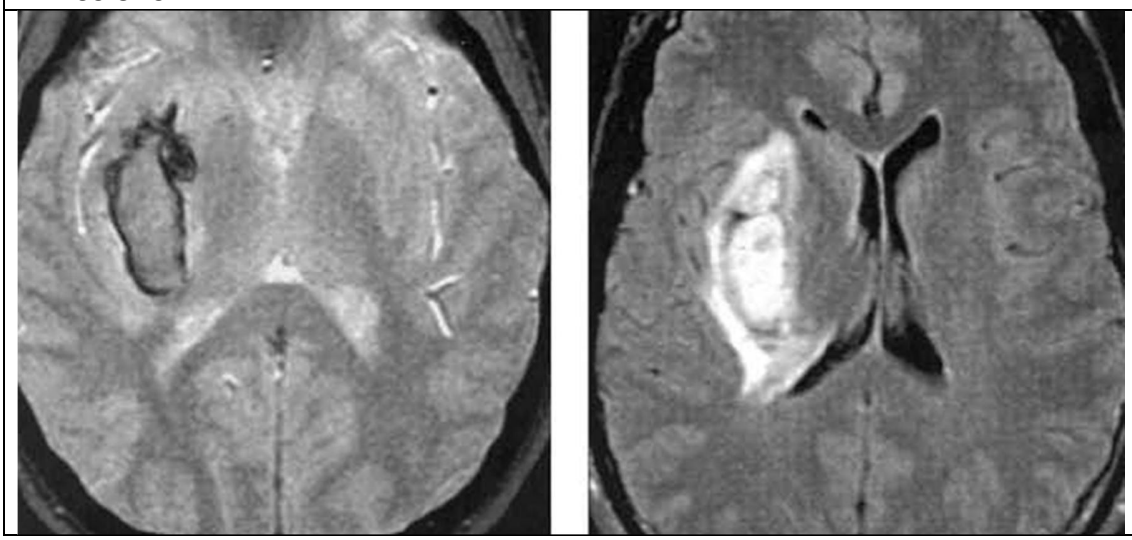
1) Heroin Inhalation:

- CT: Hypodensity
- T2: Hyperintense
- Location: Involvement of
  - Cerebellar WM
  - Posterior cerebral WM
  - Posterior limb of internal capsule.
  - Toxic leukoencephalopathy
  - Sparing of subcortical WM & dentate nuclei



2) Cocaine:

- Hemorrhagic (80%) > ischemic stroke
- CT: ICH, SAH, IVH
- T2: severe hyperintense lesions of cerebral and insular subcortex WM lesions



VII- CNS Complications of Radiation Therapy

1) Leukoencephalopathy
2) Parenchymal volume loss

3) Mineralizing microangiopathy
4) Cryptic malformations

5) Radiation Necrosis.
6) Second Neoplasms.

7) Neuroendocrine Pathology
8) Neuropsychological Delay

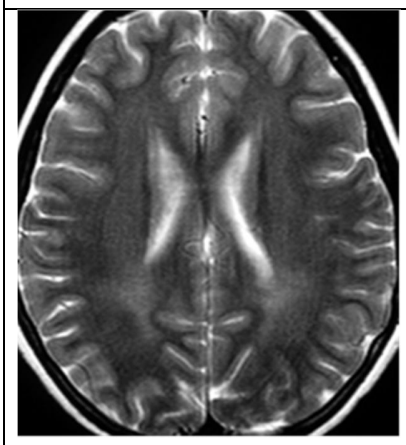
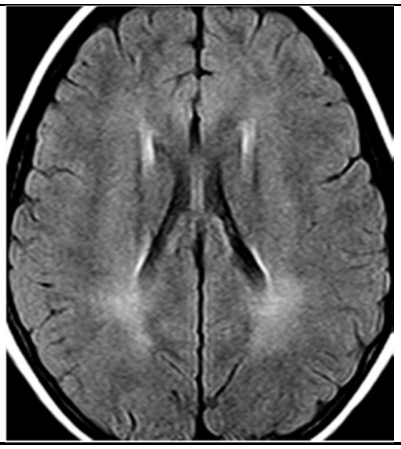
1) Radiation Leukoencephalopathy

Neurotoxic reaction to radiation therapy divided into:

- Acute injury (1-6w): Mild and reversible, vasogenic edema
- Early delayed injury (6w-months): Edema and demyelination
- Late delayed injury (months-years): more severe and irreversible; Necrosis and demyelination

<u>Imaging findings:</u>

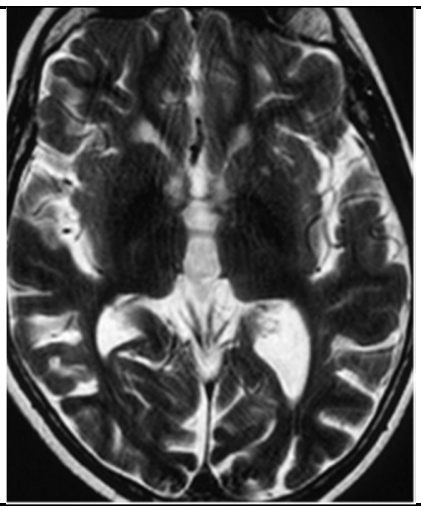
- T2 & FLAIR: hyperintense signal
- Diffuse, symmetrical involvement of central and periventricular WM
- Relative sparing of subcortical U-fibers.

Axial T2	Axial FLAIR
	

2) Parenchymal Brain Volume Loss

- Children receiving CNS prophylaxis with irradiation or intrathecal chemotherapy may develop cerebral volume loss, even when CNS leukemic involvement was not originally present.

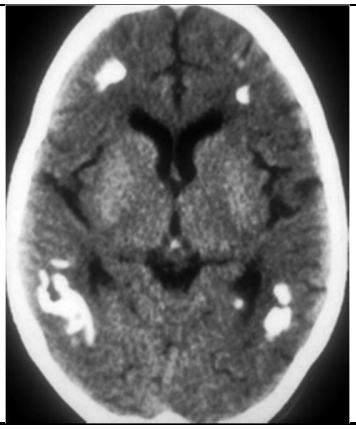
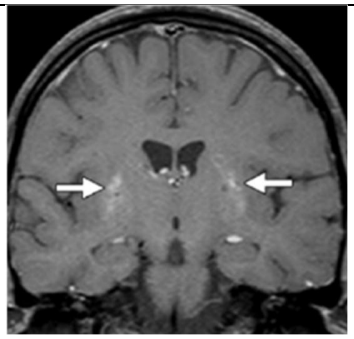
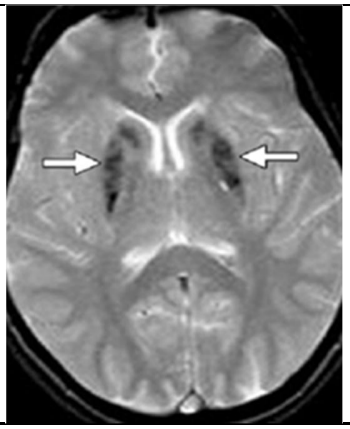
- T2/FLAIR: shows enlarged ventricles and subarachnoid spaces.



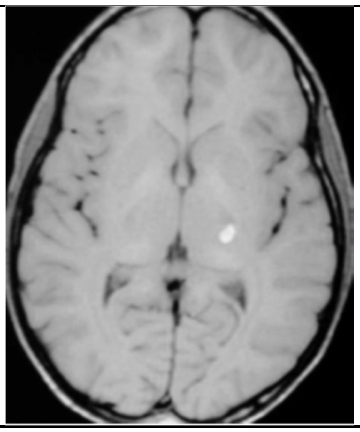
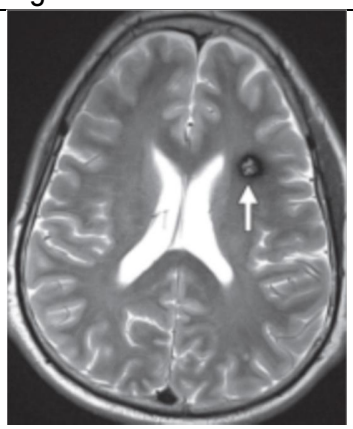
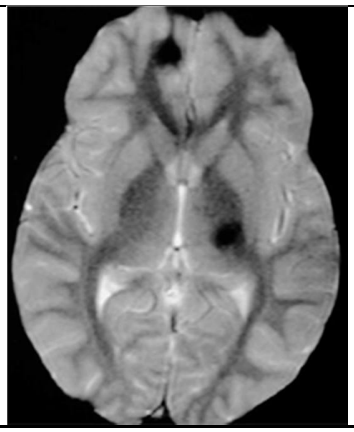
3) Mineralizing Microangiopathy

Imaging findings:

- Location: Basal ganglia and subcortical white matter

CT: hyperdense Dystrophic calcifications.	Coronal T1: Hyperintense signal	Axial GRE T2*: Hypointense signal
		

4) Cryptic Vascular Malformations

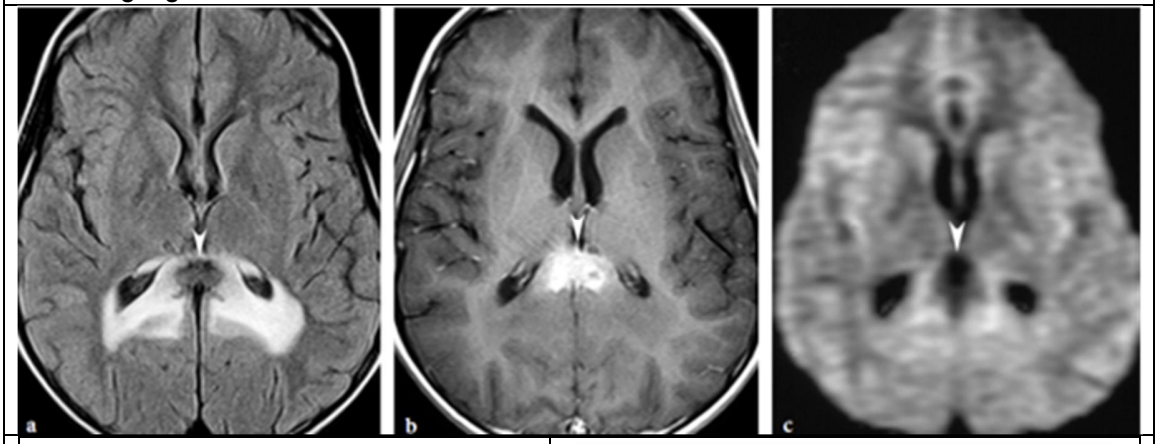
T1: high signal	T2: Popcorn surrounded by low signal hemosiderin rim	T2*: blooming artifact
		

5) Radiation Necrosis:

Histologically, focal radiation necrosis corresponds to coagulation necrosis of the white matter, accompanied by fibrinoid necrosis of the vessels in the acute phase, surrounded by neovascular proliferation in the late phase.

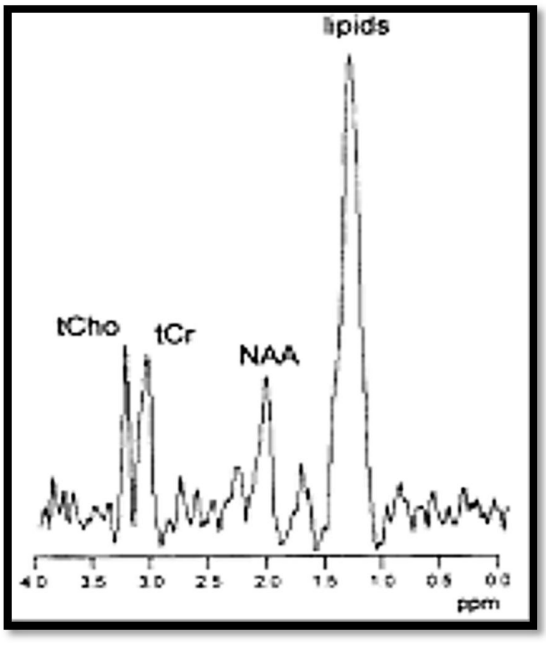
❖ Radiological findings:

- T1&T2&FLAIR: hypointense
- T1+C: Heterogeneous enhancement due to inner necrotic changes.
- DWI: No restricted diffusion.
- Surrounding marked edema
- occurring within the radiation port
- Differentiating radiation necrosis from tumor is difficult with standard imaging alone.



❖ MR Spectroscopy:

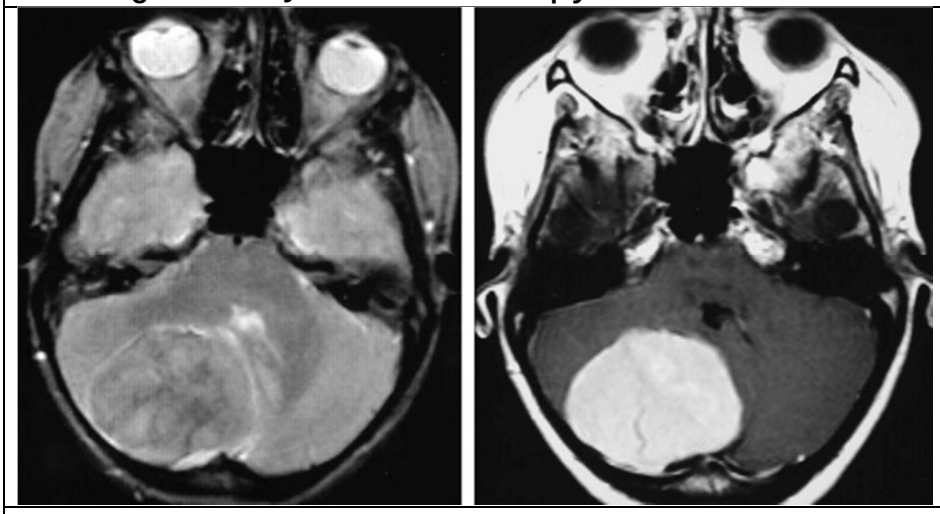
- NAA: Diminished
- Lipid+Lactate peak: indicating necrosis
- Cho:Low
- Cho/NAA,Cho/Cr: Low



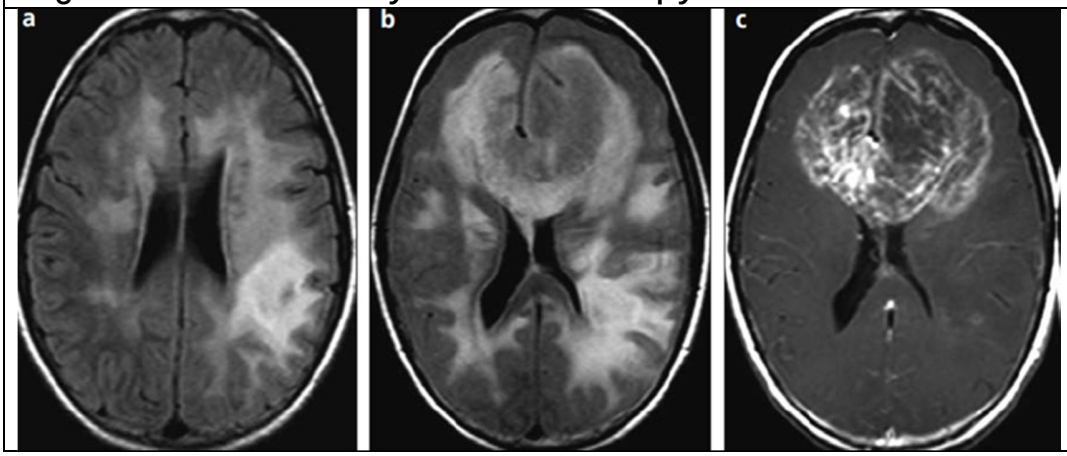
6) Secondary Neoplasm

- The most common 2nd malignant neoplasms are CNS tumors include:
  - Meningioma, sarcoma, glioma (often high grade), ependymoma, and, more rarely, primitive neuroectodermal tumor.

Meningioma 2ry to Radiotherapy



High Grade Glioma 2ry to Radiotherapy



7) Neuroendocrine Pathology:

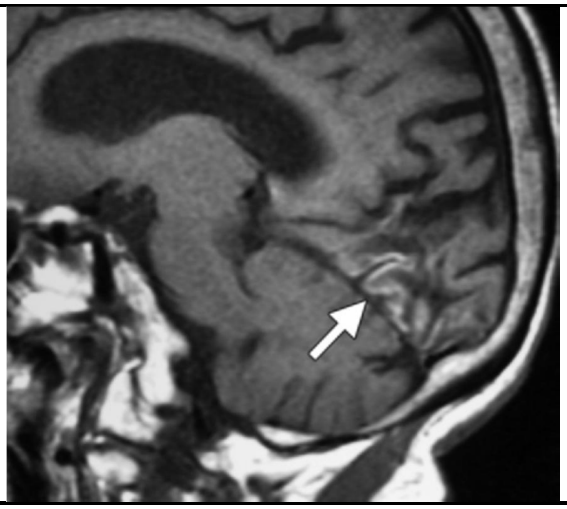
- Radiation injury to the hypothalamus-hypophyseal axis has only rarely been commented upon in the radiological literature.
- Laboratory tests may reveal hormonal alterations as early as 9–12 months after treatment, whereas clinical manifestations may not become apparent before years.
- Therefore, laboratory surveillance is crucial in order to administer replacement treatment before definitive sequelae, such as short stature, develop.

8) Neuropsychological Delay

- Reduced intellectual quotient (IQ), learning and memory disturbances, and deficit of fine motor performances are common long-term complications of RT.
- Neurocognitive deficits range from mild learn-ing issues to severe limitations in intellectual function and quality of life.
- Remarkably, leukemic children treated with chemotherapy alone seem to function considerably better in terms of late cognitive and academic effects.

9) Cortical Laminar Necrosis

- Cortical laminar necrosis is a sequela of a global hypoxic ischemic event or, less commonly, an effect of immunosuppressive therapy or radiotherapy or chemotherapy
- T1: High-signal intensity cortical lesions + parenchymal atrophy progresses.



VIII- CNS Complications of Chemotherapy

1) Cerebrovascular hemorrhage

2) Cerebral venous thrombosis

3) Reversible posterior encephalopathy (PRES).

4) Methotrexate leukoencephalopathy.

5) Disseminated necrotizing leukoencephalopathy (DNL)

6) Anterior lumbosacral radiculopathy.

1) Cerebrovascular Hemorrhage:

- The effect of some chemotherapeutic agents such as L-asparaginase aggravates the underlying thrombocytopenia and coagulopathy of leukemia.
- Subdural or subarachnoid extraaxial hemorrhage is less common than intraaxial.

2) Cerebral Venous Thrombosis

- Cancer is one of the most common causes of pediatric venous thromboembolism.
- ALL is the most common malignancy associated with venous thromboembolism in children.

3) Posterior Reversible Encephalopathy Syndrome (PRES)

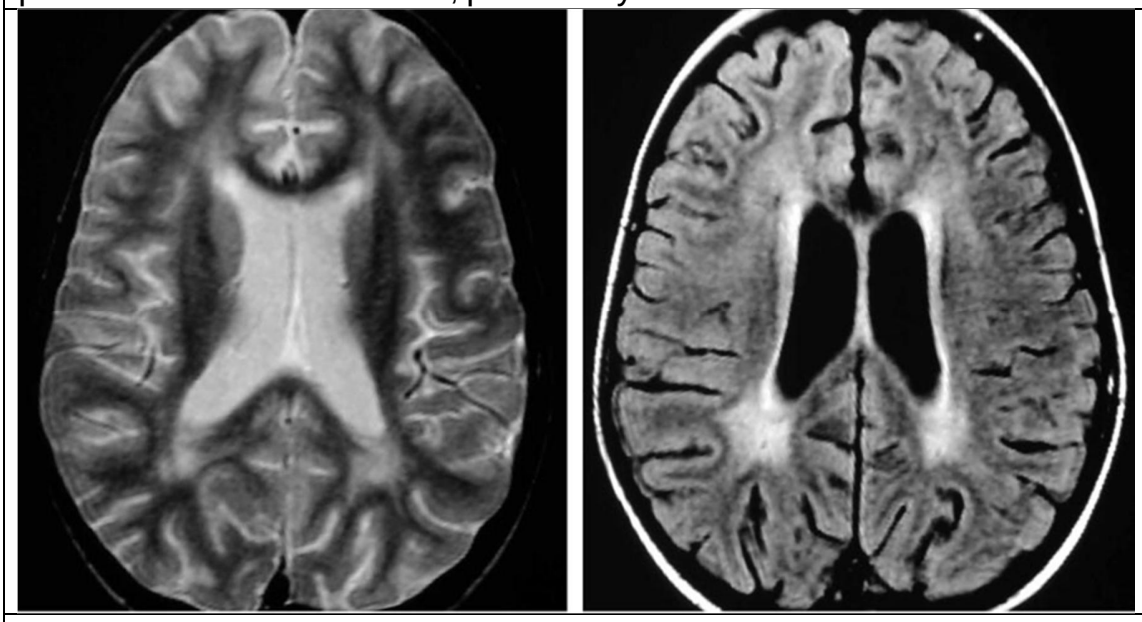
- PRES is a cliniconeuroradiologic disease
- Characteristic MR imaging findings of cortical and subcortical hyperintense signal on T2 and FLAIR
- Clinical neurologic alterations ranging from headache, altered mental status, seizures, and vision loss, to loss of consciousness.

❖ <u>Pathogenesis of PRES:</u>

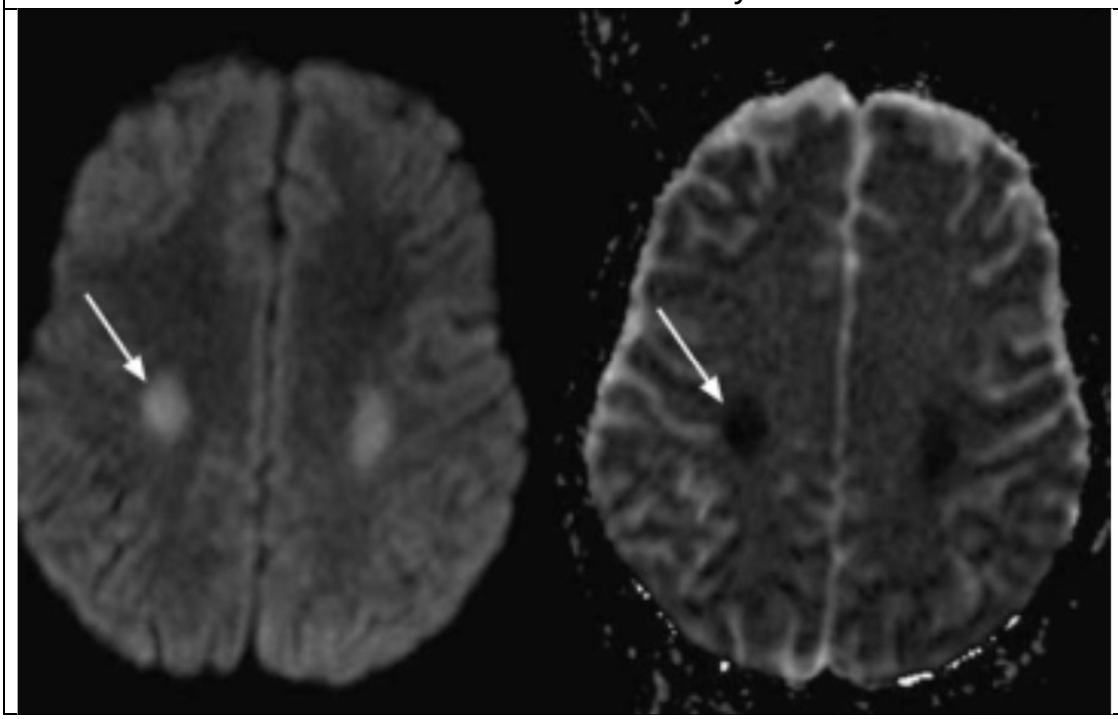
- Direct cytotoxicity of chemotherapy drugs on the cerebrovascular endothelium, with breakdown of the blood-brain barrier, leads to fluid shifts from the intravascular to the extravascular compartment caused by inadequate autoregulation of cerebral blood flow.
- Systemic hypertension: cancer patients more typically manifest only modest elevations in blood pressures

4) Methotrexate Neurotoxocity

T2/FLAIR: confluent areas of hyperintensity in the supratentorial peri-ventricular white matter, particularly the centrum semiovale

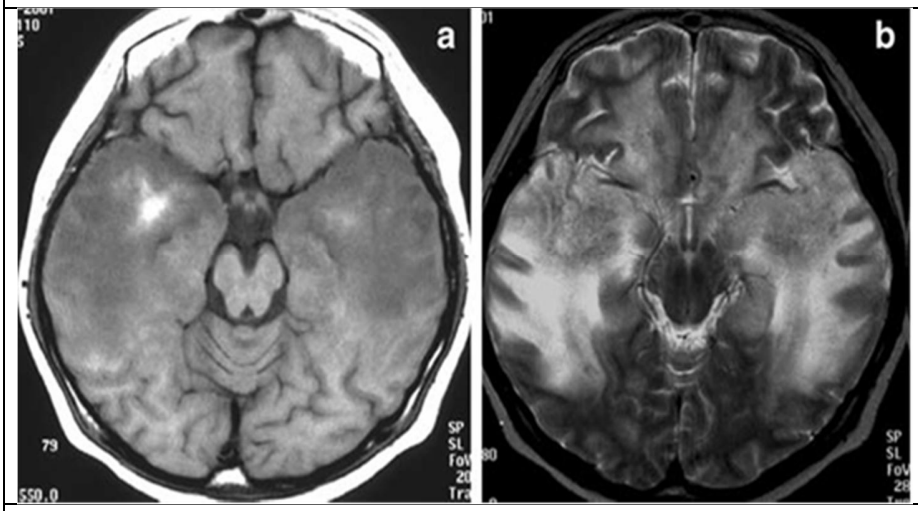


DWI: shows restricted diffusion in the centrum semiovale, which is consistent with acute methotrexate neurotoxicity.



6) Disseminated Necrotizing leukoencephalopathy (DNL)

- Diffuse white matter injury after IT methotrexate
- Clinically: Rapidly progressive deterioration
- MRI: progressive spread and coalesced of the deep white matter signal to involve the entire white matter with mass effects
- Diffuse necrotizing foci: multiple low-signal foci on T2 with contrast enhancement



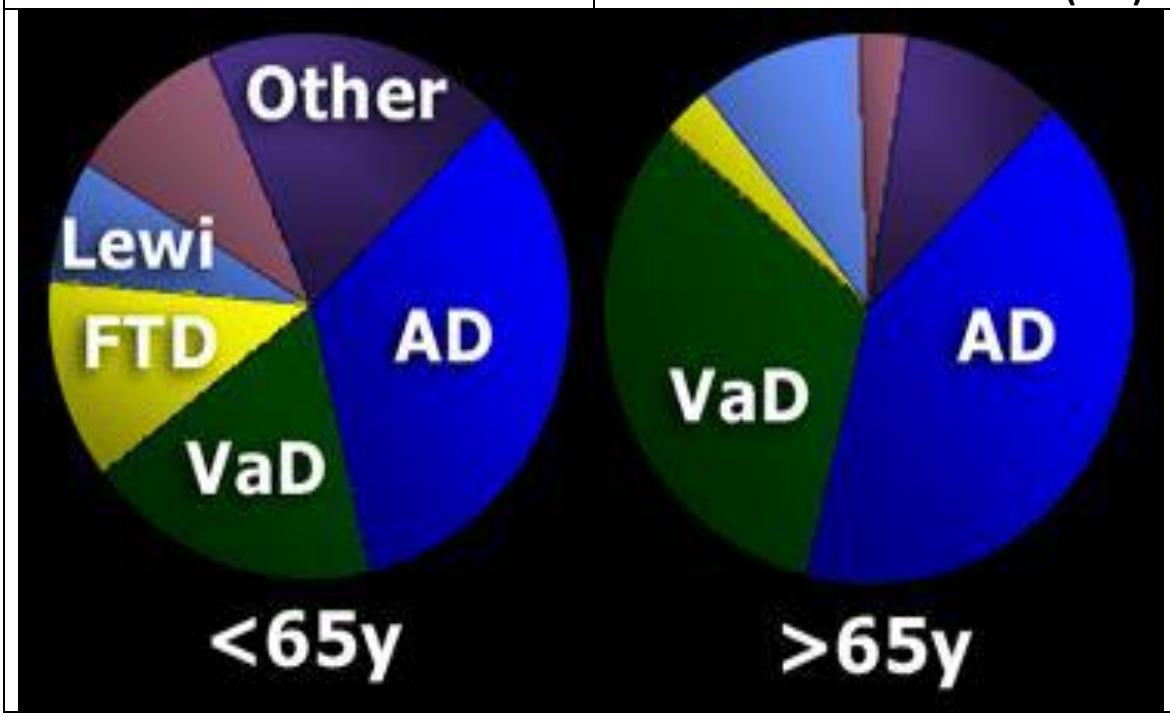
5) Anterior Lumbosacral Radiculopathy

- ALL patients undergoing treatment with IT-MTX develope progressive paraparesis.
- Sagittal and axial contrast-enhanced T1 of the spine show selective enhancement of the anterior lumbosacral roots (arrows).

Neuro Degenerative

&

Dementia Disorders

1. Aging Brain, Normal 2. Alzheimer Dementia (AD) 3. Vascular Dementia (VaD) a. Multi infarct dementia b. Binswagner's disease 4. Frontotemporal lobar Dementia (Pick's Disease) (FTLD) 5. Neurodegeneration with Brain Iron Accumulation (NBIA) ○ PKAN = Hallervorden-Spatz Syndrome: 2. Fahr Disease	3. Parkinsonian Syndromes: a. Parkinson Disease b. DLB = Dementia With Lewy Bodies c. MSA = Multiple System Atrophy d. PSP = Progressive Supranuclear Palsy. e. CBD = Corticobasal Degeneration 4. Amyotrophic Lateral Sclerosis (ALS) 5. Wallerian Degeneration 6. Creutzfeldt-Jakob Disease (CJD)
	

1) Normal Aging Brain

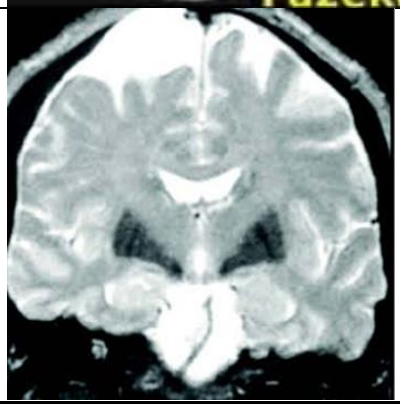
↓ Overall brain volume
Relative ↑ CSF spaces

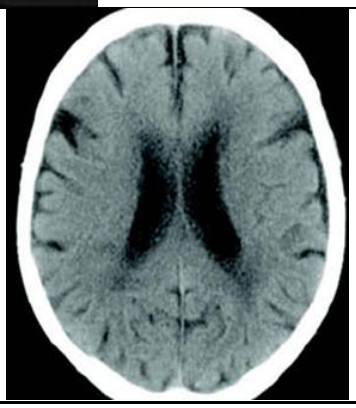
Imaging findings:

- Widened cortical sulci
- Enlarged ventricles,
- Patchy/confluent periventricular rim of low CT density / high T2 & FLAIR
- ± Symmetrical, punctate calcifications in globi pallidi (GP)

- **Lacunar Infarcts:**
  - "Infarct-like" T2 hyperintense lesions
  - 70% < l0 mm
  - Mostly in basal ganglia (BG), thalami

- **Normal Iron Concentration = Low signal on T1 & T2**
  - In normal adults: Areas of maximum iron concentration are found in: GP, Red nucleus, substantia nigra, Dentate nucleus
  - With aging (8th decade): iron concentration increase in caudate and putamen and may equal to GP





2) ALZHEIMER DEMENTIA

- **1st common cause** of Neurodegenerative dementia (50-75%)
- Progressive degenerative disease of brain due to abnormal accumulation of tau protein, which plays a key role in neuronal/glial dysfunction and cell death.

Risk factors:

- Age is a strong risk factor:
    - 8% of individuals over the age of 65 y
    - 30% of individuals over the age of 85 y advanced age
- Female gender
- apolipoprotein E (APOE) ε4 allele carrier status
- Current smoking
- Family history of dementia
- Mutations of amyloid precursor protein
- Down syndrome

Imaging findings:

- Preferential volume loss in temporal & parietal lobes
- Atrophy of hippocampus and parahippocampal gyrus
- ↑ CSF spaces surrounding medial temporal lobes
- ↑ Perihippocampal fissures
- ↑ Temporal horns

MR Spectroscopy:

- ↓NAA, ↑myoinositol, ↑ phosphomonoester in medial temporal and, to lesser degree, parietal lobes
- Useful to monitor progression of disease by measuring progressive ↓NAA

Nuclear medicine (SPECT and PET)

- Detect regional hypoperfusion / hypometabolism in a bi-parietal and bi-temporal distribution

F-18 FDG PET

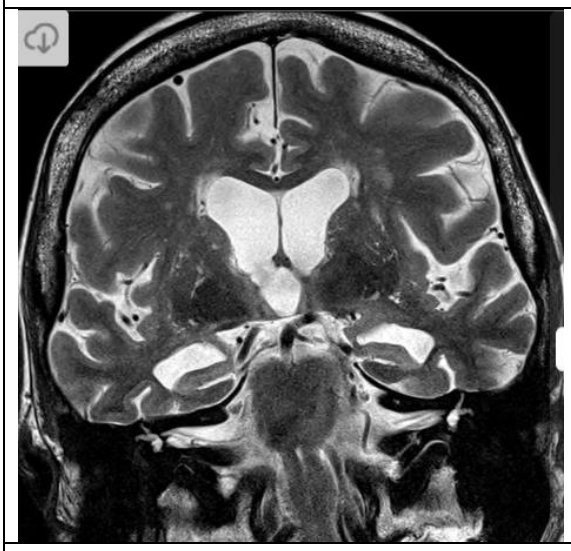
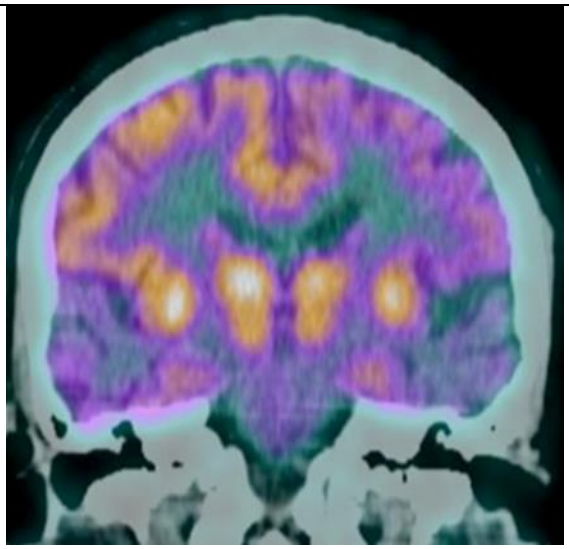
- Shows bilateral tempro-parietal, precuneus and posterior cingulate hypometabolism which is usually symmetric.
- Frontal lobes may be involved in late stage

Amyloid PET

- With increased cerebral amyloid-β (Aβ) deposition, increased activity is demonstrated in the cortex.
- It is particularly useful in excluding Alzheimer's disease as the cause of dementia, as a negative amyloid PET scan makes the diagnosis unlikely

Tau PET

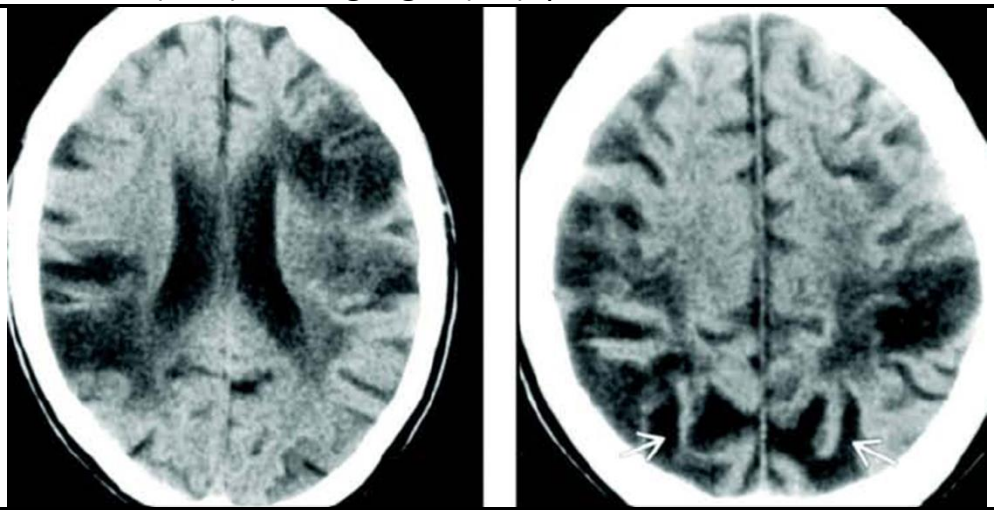
- Newer PET agents that bind to tau proteins are being investigated, which result in increased activity in the expected locations (hippocampus, entorhinal cortex and temporal and parietal cortex)

Coronal T2	Coronal T1
	
Coronal CT	Coronal PET CT
	

3) Vascular Dementia:

A) Multi Infarct Dementia

- **2nd most common cause** of dementia in elderly after AD
- Stepwise progressive deterioration of cognitive function secondary to repeated cerebral infarctions.
- Multifocal infarcts involving cortical gray matter, subcortical white matter (WM), basal ganglia (BG), pons.



B) Binswanger Disease

= Subcortical Arteriosclerotic Encephalopathy (SAE)

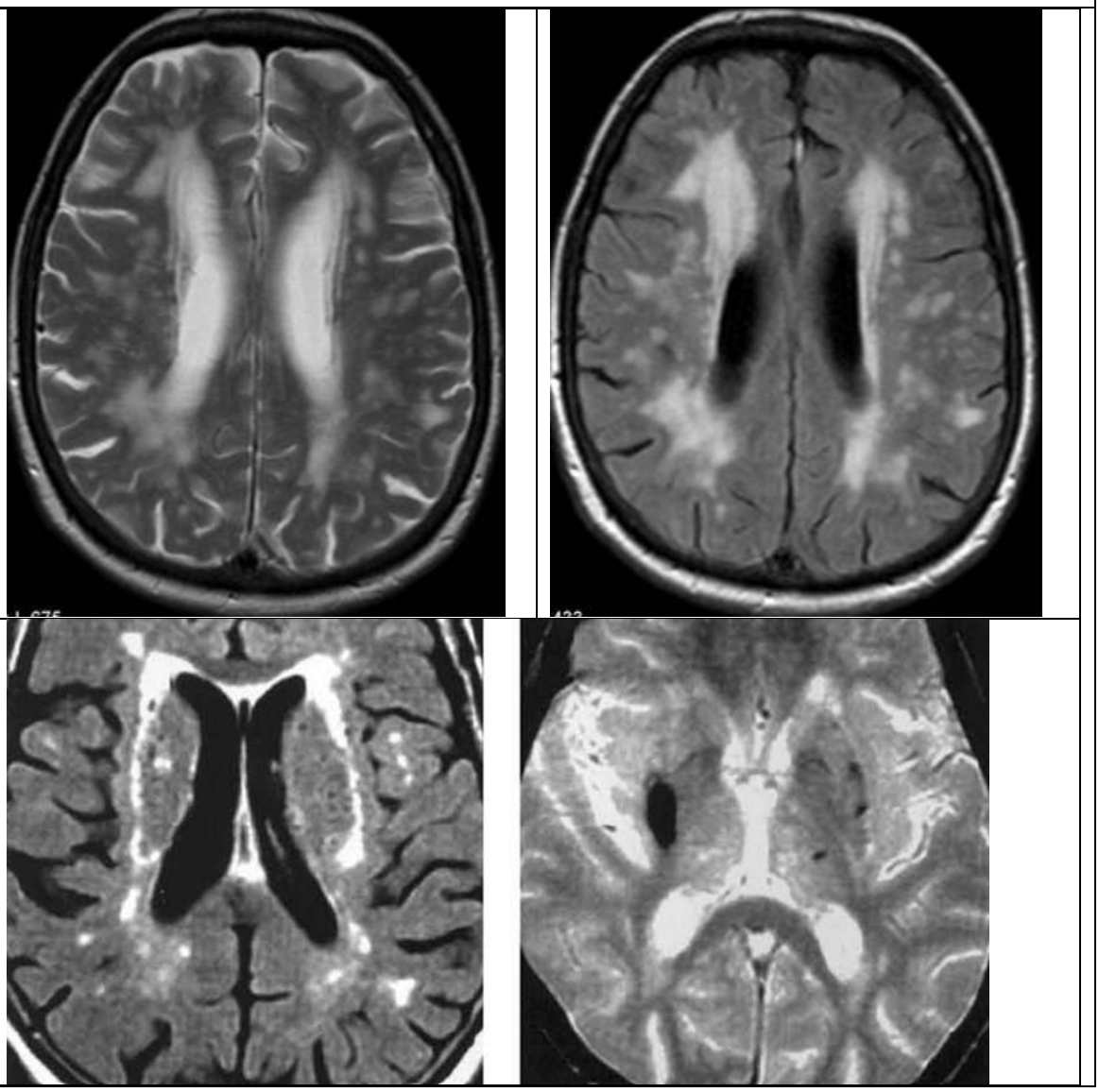
- Is form of vascular dementia
- Severe arteriosclerosis of the small penetrating arteries, which are thickened, hyalinized, stenotic, or even occluded.

Clinical Picture:

- Binswanger's disease is a clinic-pathologic diagnosis.
- Hypertension
- Dementia
- Pseudobulbar state
- Gait disorder often of Parkinsonian type,
- TIA = transient ischemic attack
- Multiple strokes

Imaging findings:

- Diffuse, confluent, periventricular hyperintense T2 / FLAIR sheets of the central white matter, especially prominent around the frontal and occipital horns and in the centrum semiovale.
- Lacunar infarcts are common in:
    - Deep central gray matter structures (BG and thalamus)
    - Corona radiata and centrum semiovale.
- Moderate cerebral atrophy.
- Petechial hemorrhage:
    - Hyperintense in T1
    - hypointense in T2*
    - Predilection for BG and thalami

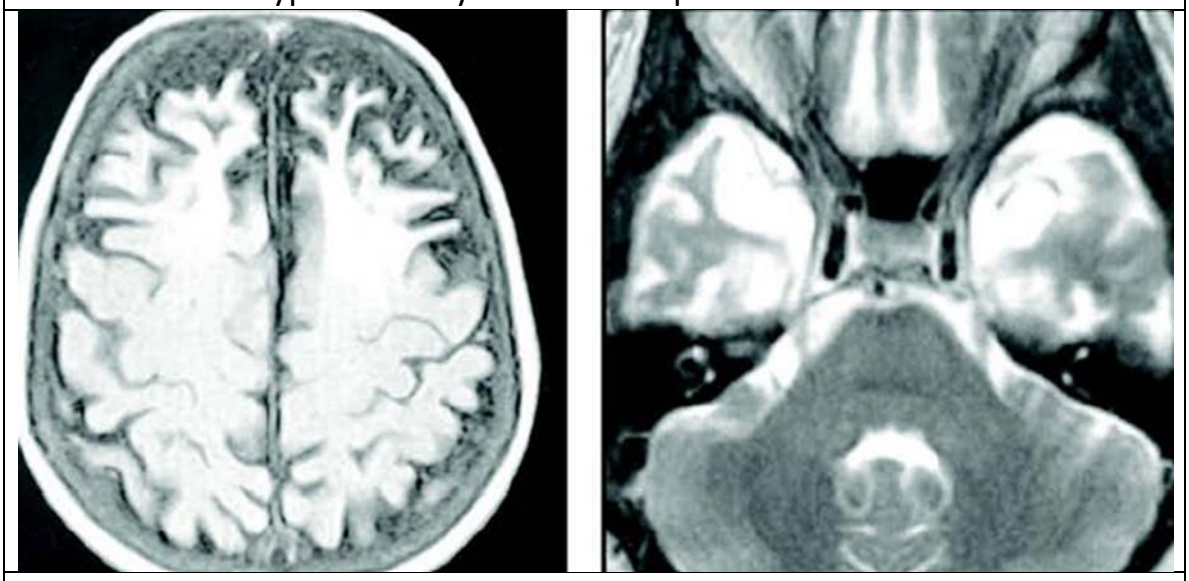


4) FRONTOTEMPORAL LOBAR DEMENTIA (FTLD) = Pick disease

- Accounts for 5-10% of cases of dementia.
- Clinically characterized by behavioral and language disturbances that may precede or overshadow memory deficits.
- No treatment for this condition.

Imaging findings:

- Anterior frontal + temporal atrophy:
  - Gyral atrophy
  - Sulcal enlargement
  - Ventricular enlargement
- T2 & FLAIR: Hyperintensity in frontotemporal white matter



5) Neurodegeneration with Brain Iron Accumulation (NBIA)

- NBIA is a heterogeneous group of disorders characterized by brain degeneration and excessive iron deposition in the basal ganglia.
- Two clinical categories are recognized:
  - Classic early-onset, rapidly progressive disease.
  - Atypical late onset, slowly progressive disease.
- In classic NBIA and in one-third of cases of atypical disease, the PANK2 gene is mutated. This gene encodes a pantothenate kinase.

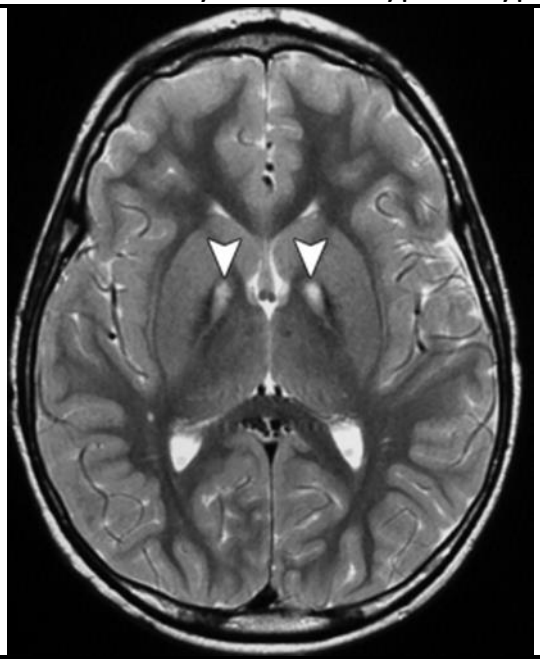
❖ Pantothenate kinase-associated neurodegeneration (PKAN) = Hallervorden-Spatz Syndrome:

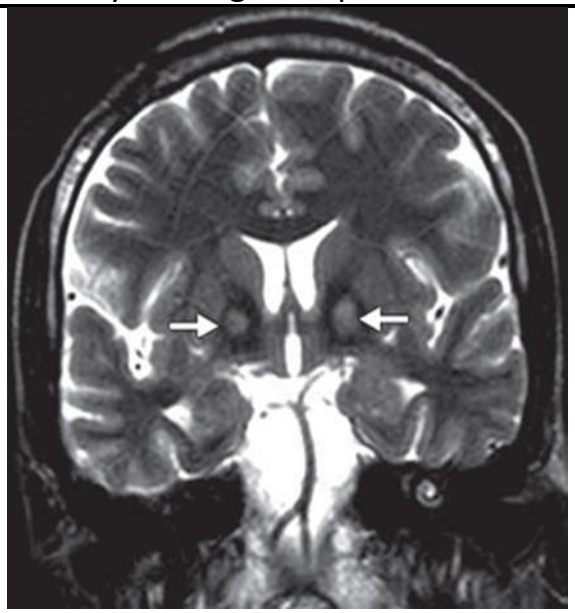
- It is a subset of neurodegeneration with brain iron accumulation (NBIA).
- It is an autosomal recessive disorder causing involuntary spasticity and progressive dementia.

Imaging findings:

T2 WI: bilateral hypointensity in the globus pallidus, which correlates with iron accumulation.

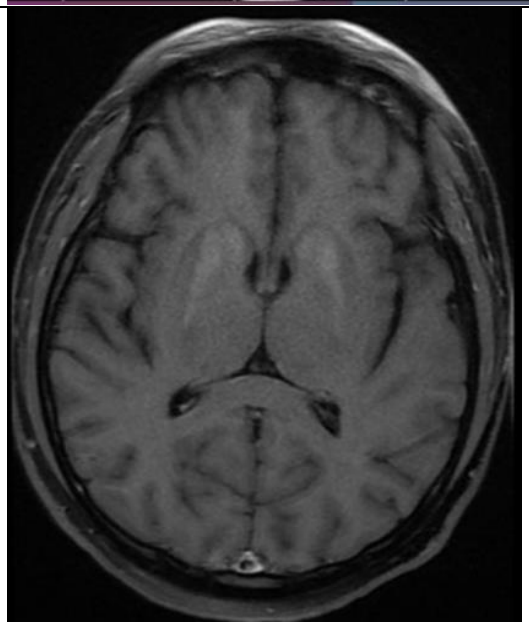
T2 WI: "**eye of the tiger sign**" with a high-signal-intensity center surrounded by the more typical hypointensity in the globus pallidus.





6) Fahr disease

- A rare neurodegenerative disease
- Bilaterally symmetric calcification in:
    - Basal ganglia
    - Thalamus
    - Dentate nuclei
    - Centrum semiovale in the absence of hypoparathyroidism
    - Subcortical white matter of the cerebrum



7) Parkinsonian Syndromes

a. **Parkinson Disease**
b. **DLB = Dementia With Lewy Bodies**
c. **MSA = Multiple System Atrophy**
d. **PSP = Progressive Supranuclear Palsy.**
e. **CBD = Corticobasal Degeneration**

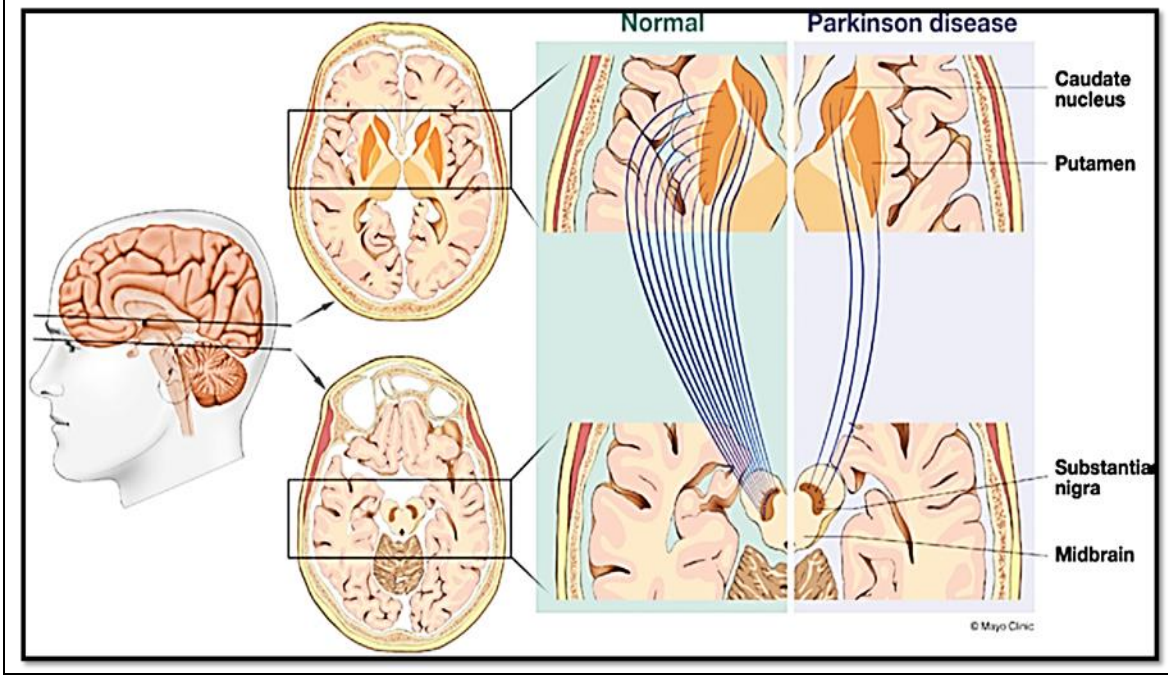
7-A) Parkinson Disease

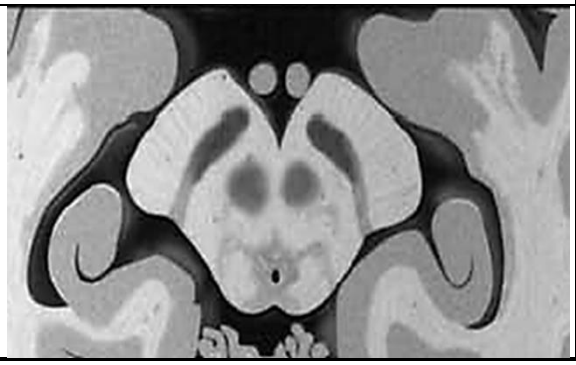
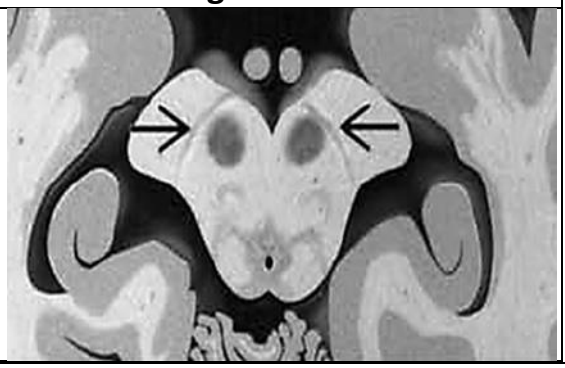
- Progressive neurodegenerative disease predominantly caused by primary disorder of pars compacta of substantia nigra (SN).
- Location: SN + Putamen

Substantia Nigra (SN):

SN= 2 layered gray matter (GM) structure on axial T2WI at upper midbrain level in normal subjects:

- Pars Reticulata Of SN (Snpr): Hypointense area in posterior region of crus cerebri
- Pars Compacta Of SN (Snpc): Relatively hyperintense area between SNPr, red nucleus (RN)



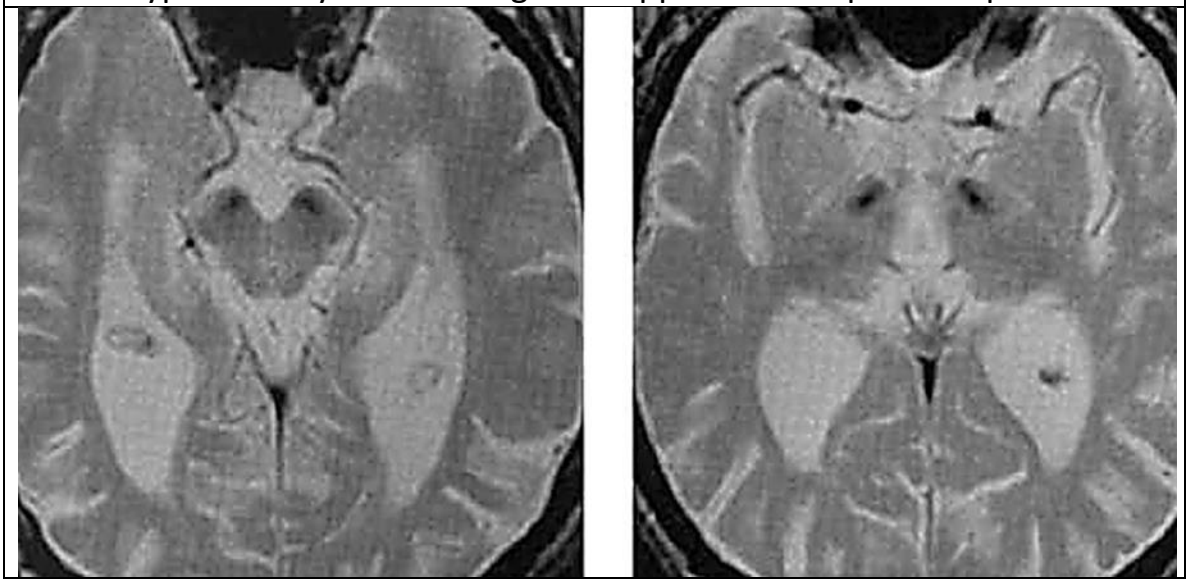
Normal SN Anatomy	**Parkinson disease** **Narrowing and depigmentation of substantia nigra**
	

Clinical Picture:

- Bradykinesia
- Tremor
- Rigidity
- Gait disturbance
- Excellent initial response to dopaminergic therapy

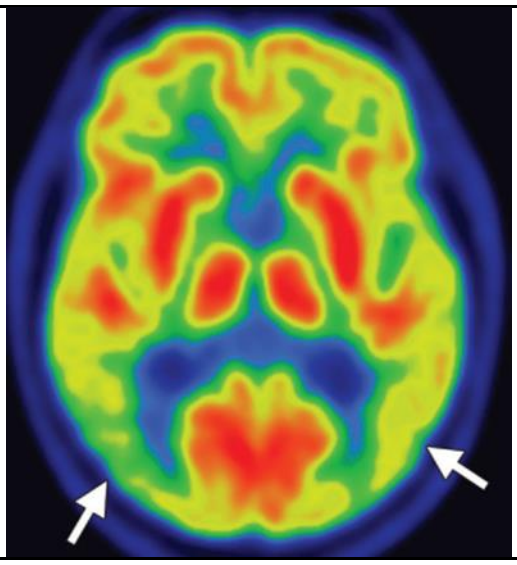
Imaging findings:

- T2: Hypointensity + Narrowing or disappearance of pars compacta of SN.



FDG PET

- Normal
- Preserved metabolism in the basal ganglia (putamen and globus pallidus)
- DD: PSP and MSA => reduced basal ganglia FDG activity.



7-B) Dementia with Lewy bodies (DLB) = Lewy body disease

- 25% of dementias
- Belongs to the atypical Parkinson syndromes
- Older patients (onset typically in 50-70 years of age), and is sporadic
- Patients typically present with one of three symptom complexes: detailed visual hallucinations, Parkinson like symptoms and fluctuations in alertness and attention.
- Pathologically: the disease is characterized by the presence of Lewy bodies in various regions of the hippocampal complex, subcortical nuclei and neocortex with a variable number of diffuse amyloid plaques.
- Cholinesterase inhibitors are currently the treatment of choice for this condition.

Imaging findings:

- Generalized decrease in cerebral volume most marked in frontal lobes
- Enlargement of the lateral ventricles
- No specific abnormalities.
- Normal hippocampus (DD: Alzheimer's disease)

7-C) Multiple System Atrophy

MSA is a neurodegenerative disease

2 forms:

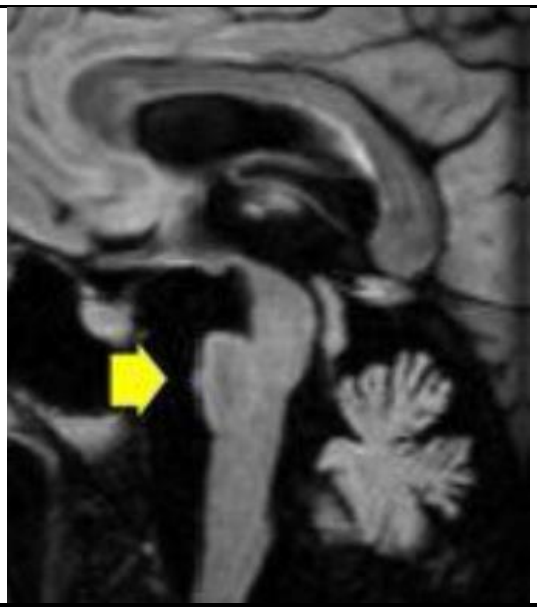
- **MSA-C:** predominance of cerebellar symptoms (olivopontocerebellar atrophy)
- **MSA-P:** predominance of parkinsonian signs and symptoms (Substania nigra degeneration)

3 Clinical presentation:

1) Shy-Drager syndrome is used when autonomic symptoms predominate
2) Striatonigral degeneration shows predominant parkinsonian features
3) Olivopontocerebellar atrophy demonstrates primarily cerebellar dysfunction

MSA-C: Decreased FDG activity + T2 hyperintensity of:

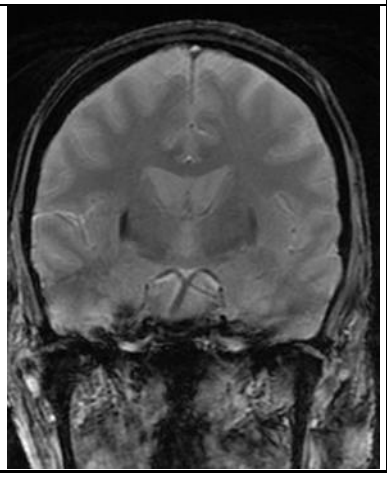
- Typically present in the pontocerebellar tracts
- Pons : hot cross bun sign
- Middle cerebellar peduncles
- Cerebellum
- Sparring corticospinal tracts

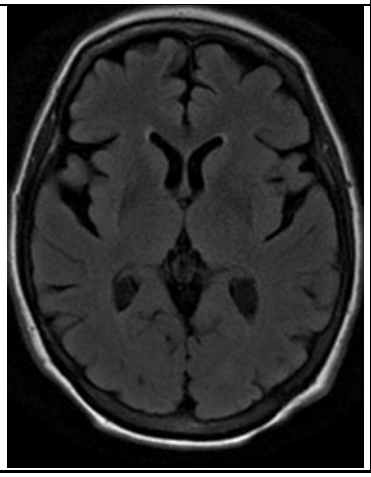


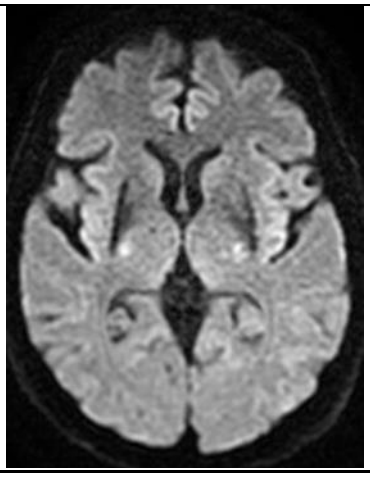
MSA-P: Putaminal findings:

- Reduced volume
- Hypointense T2 and T2* + “slitlike” marginal T2 hyperintensity
- FDG PET: Symmetric decreased FDG activity

Coronal T2	Axial FLAIR	Axial DWI

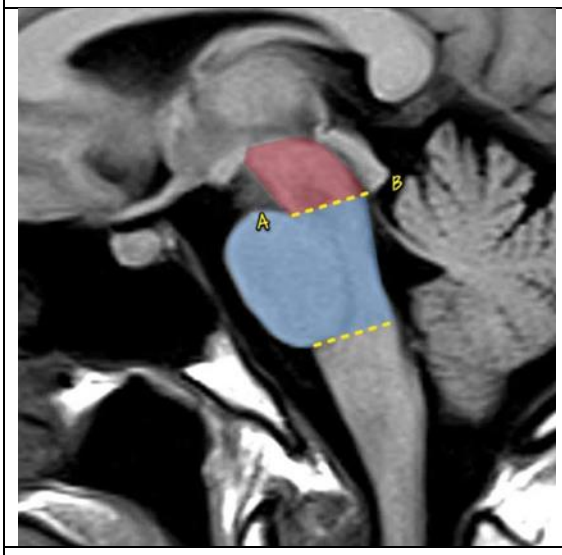
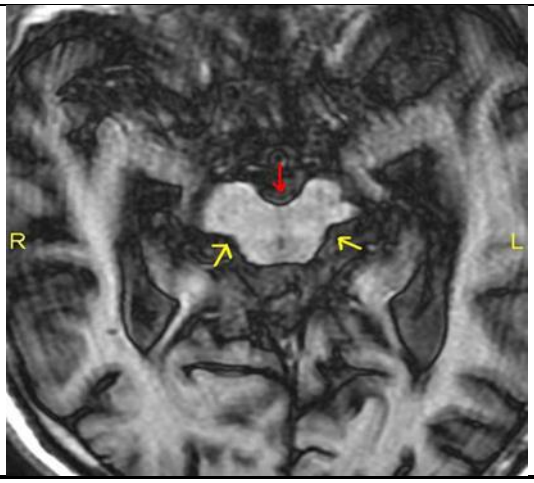






7-D) Progressive Supranuclear Palsy (PSP)

- PSP is characterized by parkinsonism with bradykinesia and rigidity, postural instability, and a pseudobulbar syndrome with dysarthria and dysphagia.
- The key feature of PSP is supranuclear palsy of vertical gaze

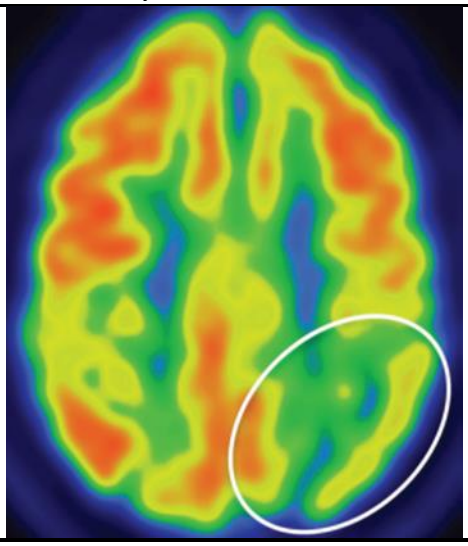
Midsagittal: Reduced midbrain to pons area ratio: approx 0.12 (Normal approx 0.24)	Sagittal: Flattening or concavity of the superior aspect of the midbrain => Hummingbird sign or "Penguin sign"
	
Axial image: • Atrophy of the midbrain and tegmentum • Reduced midbrain AP diameter at the level of the superior colliculi (from interpeduncular fossa, to the intercolicular groove <12mm • => Mickey Mouse sign	

7-D) Corticobasal Degeneration

- CBD was originally described as an asymmetric, akinetic-rigid neurodegenerative syndrome with cortical dysfunction

FDG-PET:

- Asymmetric atrophy and hypometabolism centered in the posterior frontal and parietal lobes
- Atrophy of the ipsilateral cerebral peduncle



8) Creutzfeldt-Jakob Disease

- CJD is a transmissible fatal neurodegenerative disorder caused by prions (self-replicating proteinaceous infectious particles).
- Patients with CJD typically present with rapidly progressive dementia, myoclonus, and multifocal neurologic dysfunction

4 Main Subtypes:

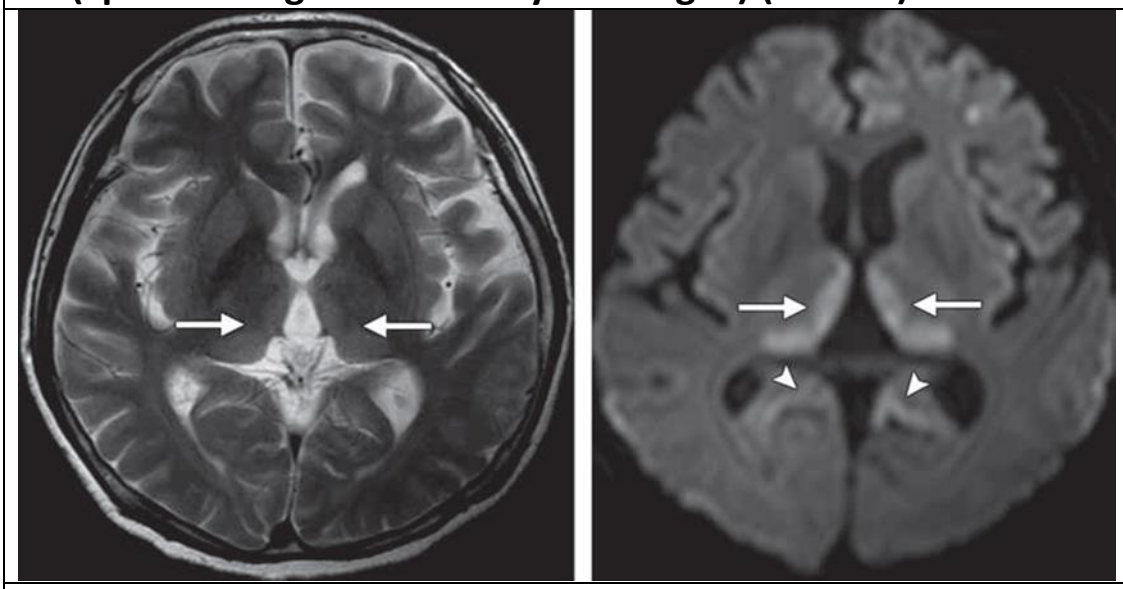
1) Sporadic
2) Familial
3) Iatrogenic
4) Variant CJD: associated with bovine spongiform encephalopathy ("mad cow disease"), transmitted through consumption of bovine spongiform encephalopathy–contaminated meat.

- Brain biopsy or autopsy: is required for a definitive diagnosis
- T2 & FLAIR & DWI: Progressive hyperintensity + Restricted diffusion of basal ganglia (Caudate & Putamen > GB), thalamus & cerebral cortex
- NO enhancement.
- EEG: Generalized periodic sharp wave complexes
- CSF: detection of 14-3-3 proteins in the CSF

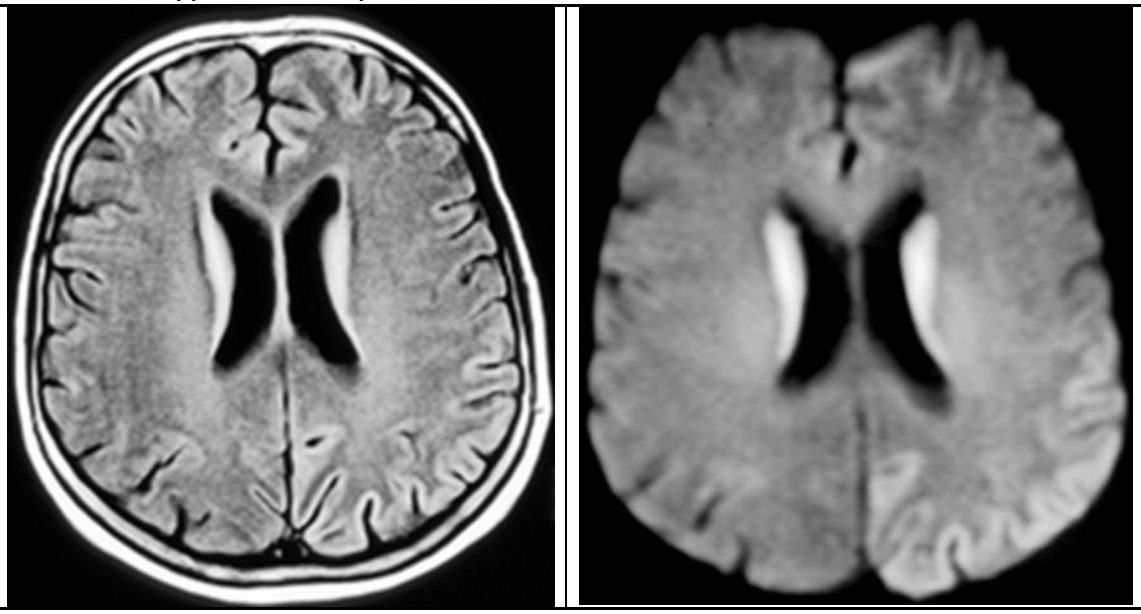
Imaging findings:

- Bilaterally symmetric areas of T2 hyperintense and restricted diffusion in the medial pulvinar nuclei of the medial thalamus

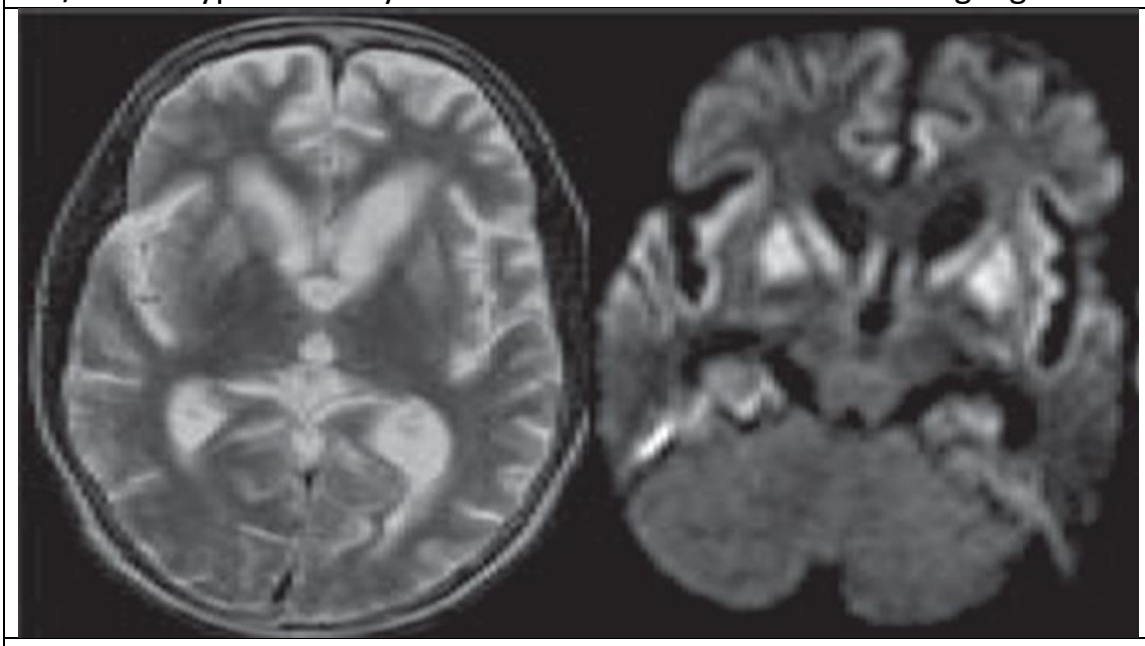
=> ("pulvinar sign" or "hockey stick sign") (arrows).



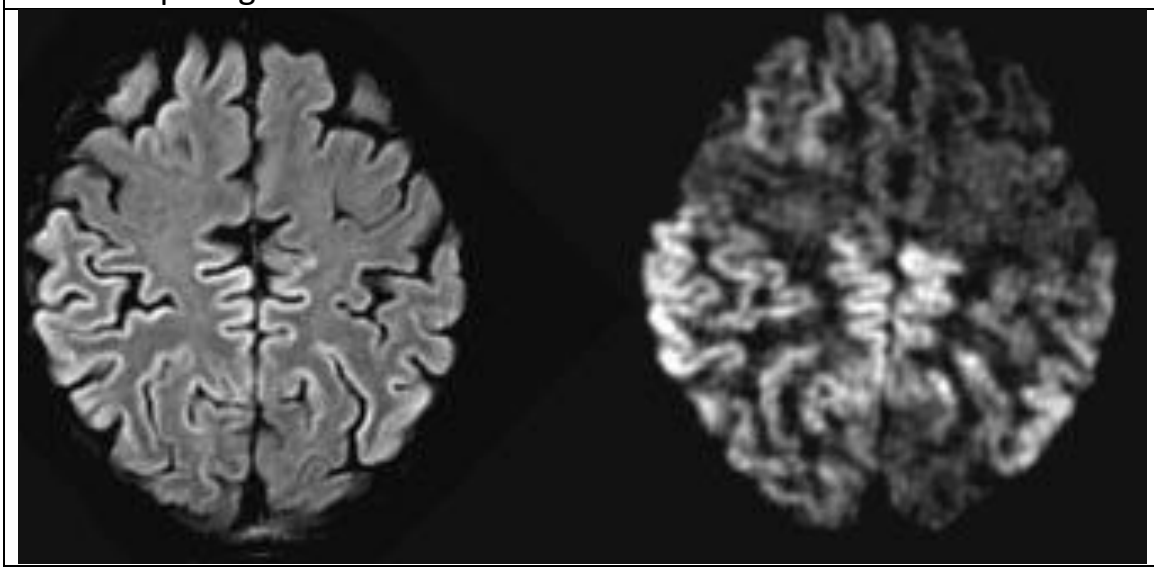
T2 / FLAIR hyperintensity and restricted diffusion of the caudate nucleus



T2 / FLAIR hyperintensity and restricted diffusion of the basal ganglia



Diffuse T2 / FLAIR hyperintensity and restricted diffusion of the cerebral cortices sparing the sensor motor cortices



9) WALLERIAN DEGENERATION

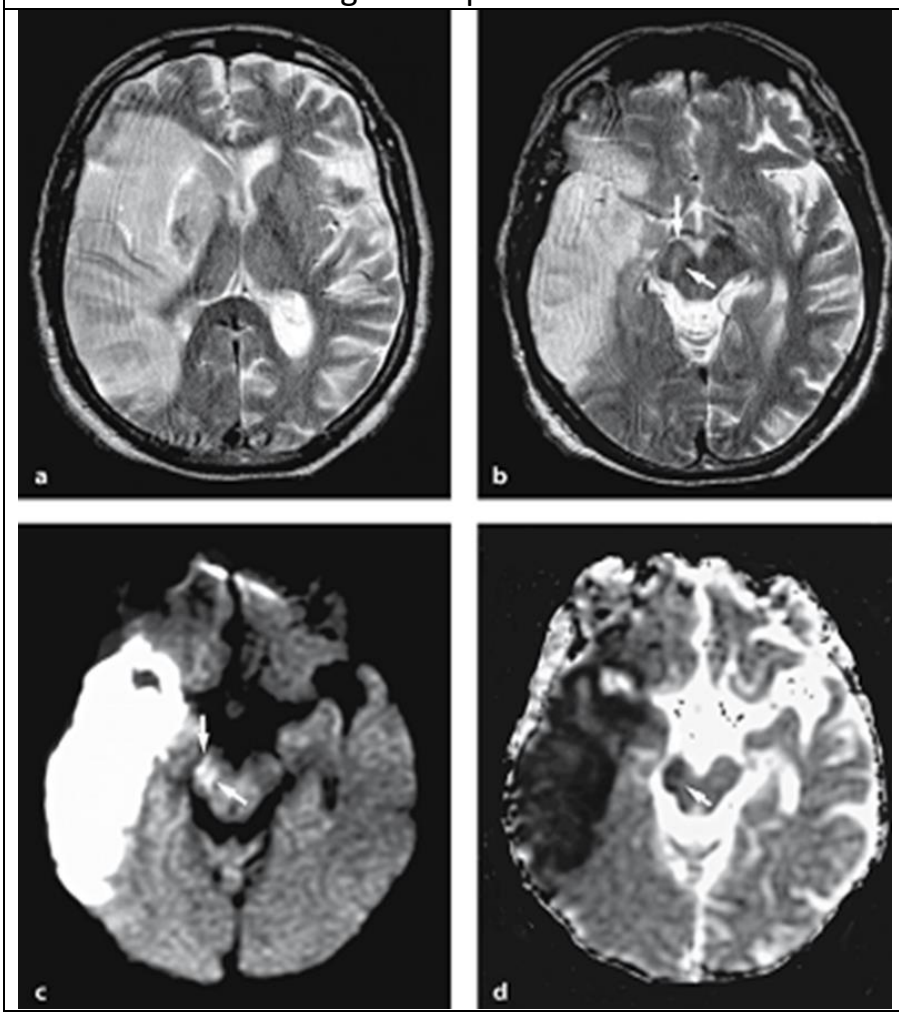
- Wallerian degeneration is an antegrade degeneration of the axons and myelin sheath resulting from injury of the proximal portion of the axons or cell bodies.
- It is most commonly recognized in the corticospinal tract secondary to MCA infarction.

Imaging Findings:

- T2/FLAIR hyperintensity and restricted diffusion within ipsilateral corticospinal tract of the territorial infarct.

Location:

- Posterior limb of IC
- Cerebral peduncles of midbrain
- Thin curvilinear regions in pons

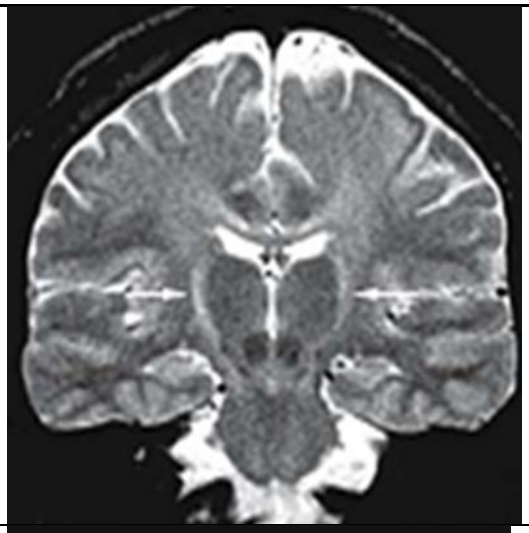
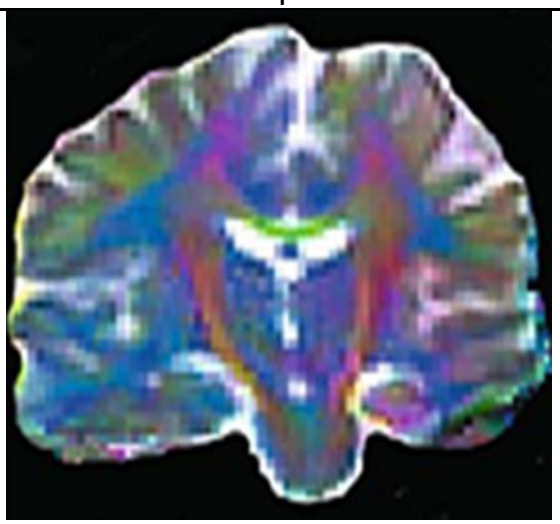
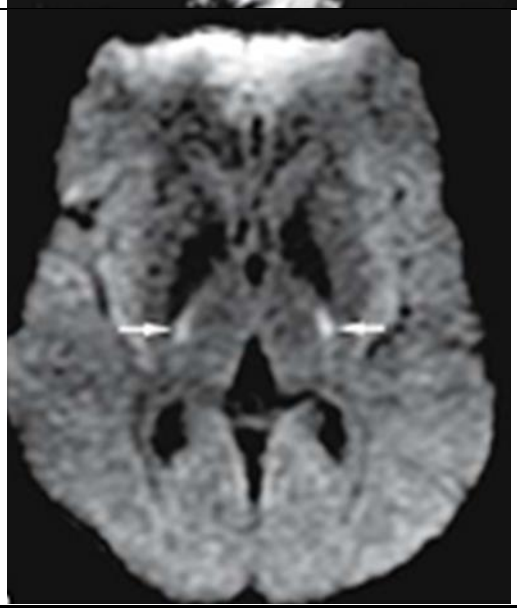
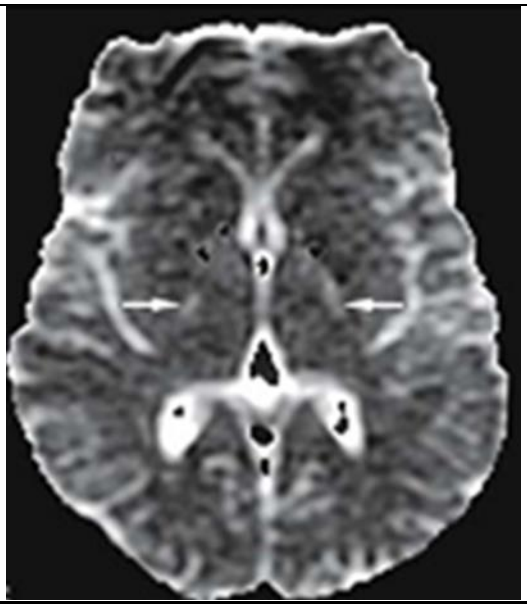


10) AMYOTROPHIC LATERAL SCLEROSIS

- Selective degeneration of:
  - Somatic motor neurons of brain stem/spinal cord (lower motor neurons (LMN))
  - Large pyramidal neurons of motor cortex (upper motor neurons, UMN)
- Amyotrophic lateral sclerosis affects middle-aged patients

Imaging findings:

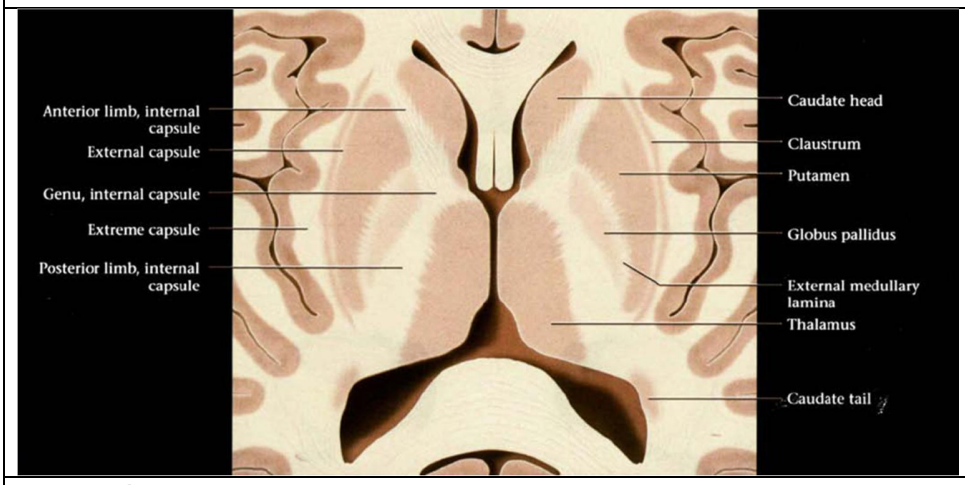
- T2/FLAIR: Bilateral hyperintensities along corticospinal tract extending from corona radiata to brainstem.
- DWI: Increased ADC and decreased FA in the corticospinal tracts.

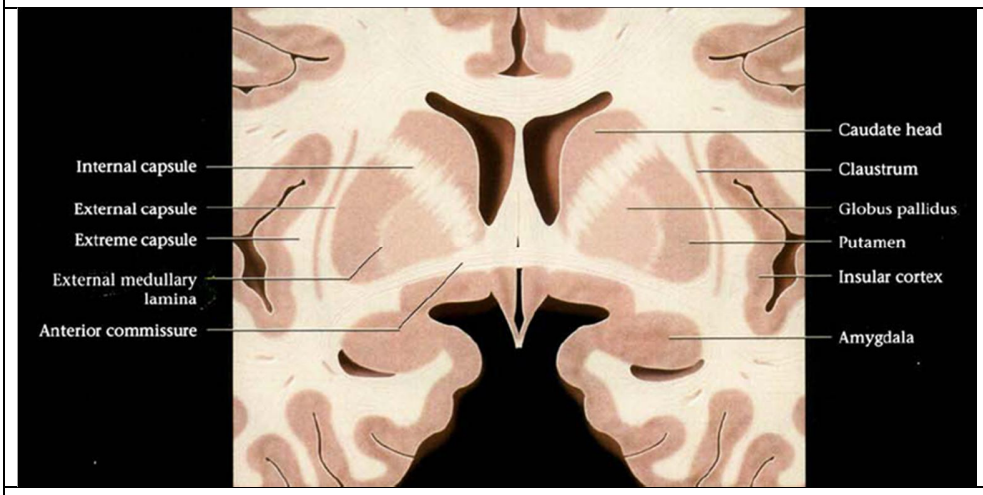
Basal Ganglia

& Thalami

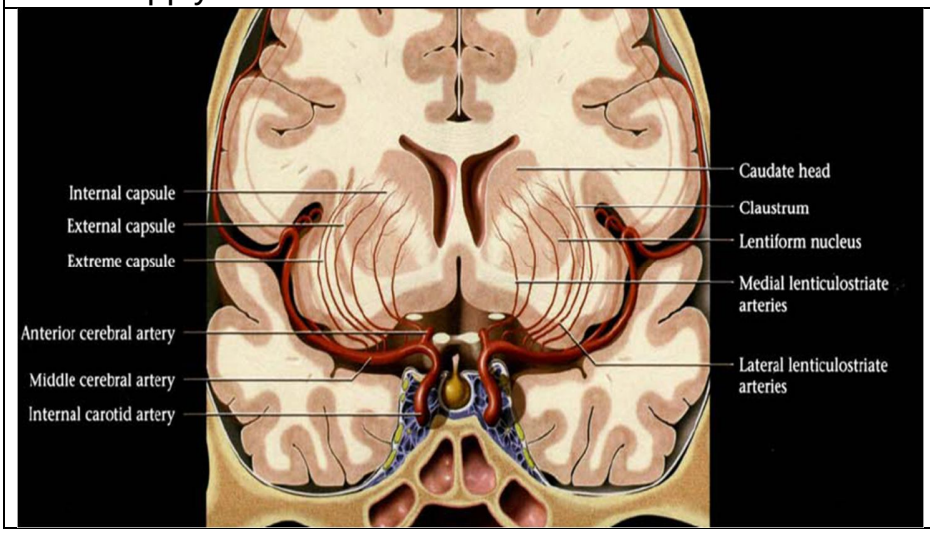
Axial



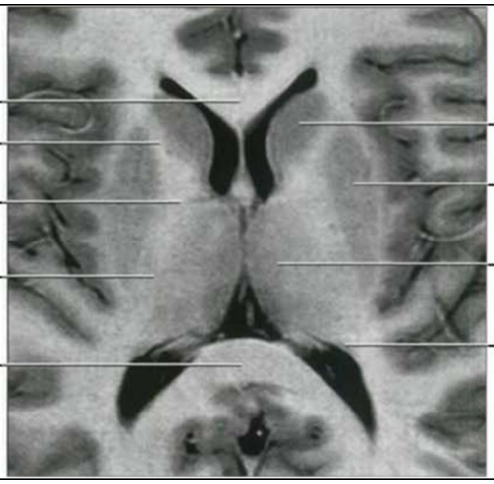
Coronal



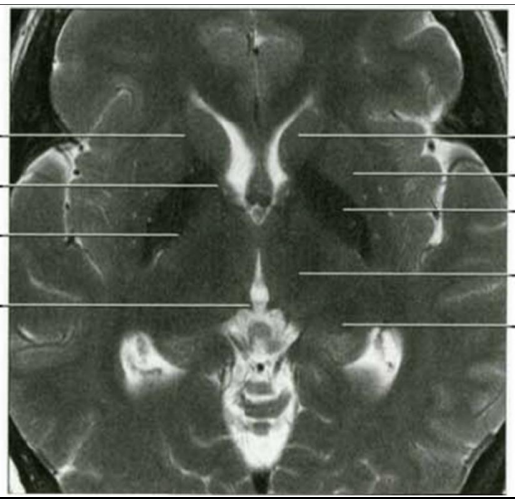
Blood Supply



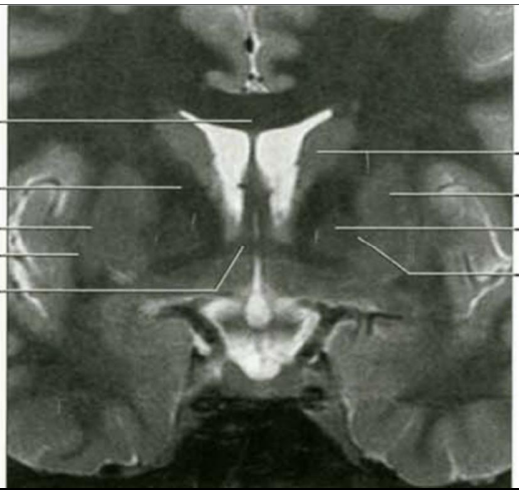
Axial T1



Axial T2



Coronal T2



❖ Bilateral BG Calcifications

- Idiopathic
  - Ageing: common, globus pallidus most commonly affected
  - Fahr disease
- Toxic
  - Carbon monoxide poisoning
  - Lead poisoning
  - Mineralising microangiopathy (Associated with radiation therapy)
- Infectious
  - TORCH
  - CNS tuberculosis
  - AIDS
  - Neurocysticercosis
  - CNS toxoplasmosis
- Metabolic
  - Parathyroid disorders
    - Hypoparathyroidism
    - Pseudohypoparathyroidism (PHP)
    - Pseudo-pseudohypoparathyroidism (PPH)
    - Hyperparathyroidism
- Inherited
  - Mitochondrial diseases e.g. MELAS
  - Cockayne syndrome
  - Pantothenate kinase-associated neurodegeneration (PKAN) (also known as hallervorden-spatz syndrome)

❖ Bilateral BG Hemorrhage

- Hemorrhagic venous infarction
- Poisoning,
- CNS Toxoplasmosis
- Japanese Encephalitis

❖ Bilateral BG Hyperintense T1 signal

- Calcifications (see before)
- Hemorrhage (see before)
- Toxins
  - Carbon monoxide
  - Hyperalimentation or long term parenteral nutrition (manganese)
- Metabolic
  - Hepatic failure
    - Acquired non-wilsonian hepatocerebral degeneration
    - Wilson's disease (copper)
  - Non-ketotic hyperglycaemic hemichorea (NHH)
  - kernicterus
- Flavavirus / Japanese encephalitis
- Hamartoma in neurofibromatosis type 1

❖ Bilateral BG Restricted DWI

- Acute exposure to respiratory chain metabolic toxins
- Carbon monoxide poisoning
- Wilson disease (early)
- Acute hepatic encephalopathy
- Hypoglycaemia
- hypoxic ischaemic encephalopathy (HIE)
- Osmotic demyelination
- Wernicke encephalopathy
- Creutzfeldt-Jakob disease
- Deep cerebral venous thrombosis
- Arterial infarction
- Flavavirus / Japanese encephalitis

❖ Bilateral BG Eye of Tiger Sign

- Hallervorden-Spatz syndrome: classical but not 100% pathognomonic
- Carbon monoxide poisoning
- Progressive supranuclear palsy
- Cortical–basal ganglionic degeneration

❖ Bilateral Thalamic

- Deep cerebral venous thrombosis
- Arterial occlusion
- Flavivirus infection
- Glioma
- Wernicke encephalopathy

❖ Bilateral BG + Thalamic + Cortical

- Hypoxia hypoxic ischaemic encephalopathy (HIE)
- Acute hepatic encephalopathy
- Hypoglycemia
- CJD
- Flavivirus infection

❖ Bilateral BG + Thalamic + Brain Stem

- Leigh Disease
- Osmotic Myelinolysis
- Wernicke encephalopathy
- Wilson disease
- Basilar Artery Occlusion

❖ Bilateral BG + Thalamic + Periaqueductal

- Wernicke encephalopathy
- Leigh Disease

❖ Bilateral BG + Thalamic + Pons

- Wilson disease
- Osmotic Myelinolysis

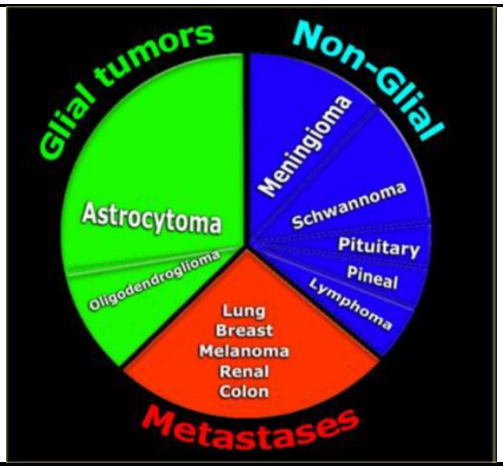
Brain Tumors

❖ Incidence

1/3 Metastases
Lung cancer is the most common

1/3 Glial tumors
Astrocytoma is the most common

1/3 Non-Glial Tumors
Meningioma is the most common



❖How To Approach Lesion by Questions ?

- **Age**
  - **Pediatric or Adult**
- **Location:**
  - **Intra- or extra-axial**
  - **Supra- or Infra-tentorial**
  - **Intra- or extra-ventricular**
- **Specific location**
- **Specific CT & MRI features**
- **Contrast enhanced MRI**
- **Mass effect**
- **Spread**
- **Grading**
  - **WHO Classification**
  - **Low grade or High grade**
  - **Advanced MRI Techniques:**
    - **MRS – DWI – Perfusion**

Q1- Age ?

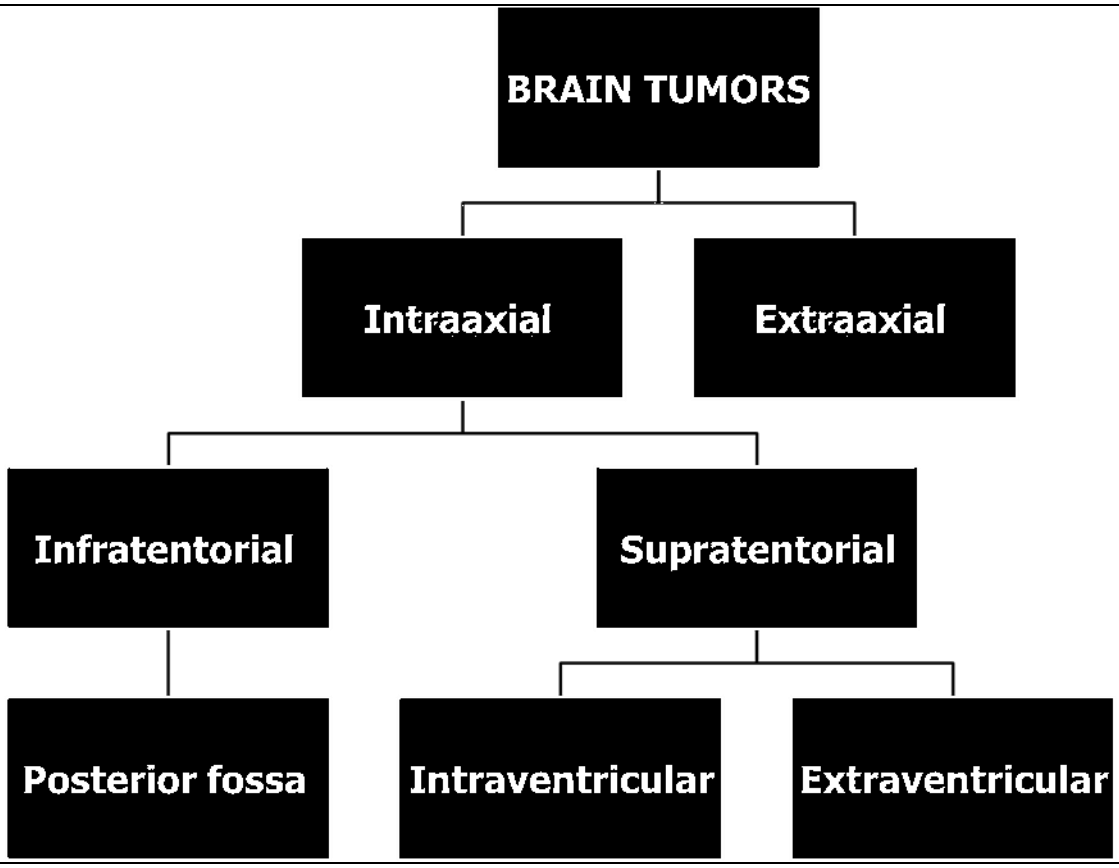
Common Intra-Axial Tumors in Pediatric

Supratentorial:	Infratentorial:
Astrocytoma	Juvenile Pilocytic Astro
Pleomorphic xanthoastro (PXA)	PNET (Medulloblastoma)
PNET	Ependymoma
DNET	Brainstem Astrocytoma
Ganglioglioma	

Common Intra-Axial Tumors in Adult

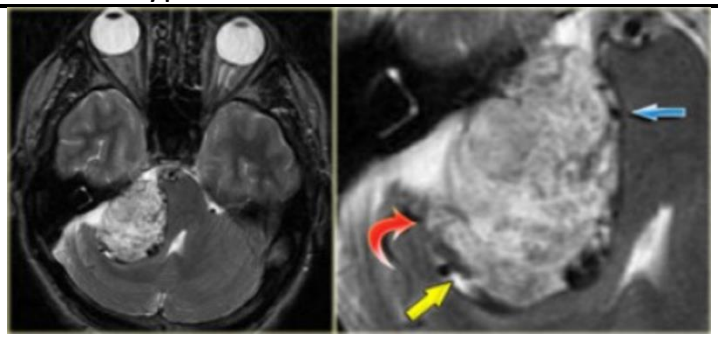
Supratentorial:	Infratentorial:
Metastases ++	**Metastases ++++**
Gliomas (25%)	Hemangioblastoma
• Fibrillary Astrocytoma	
• Anaplastic Astrocytoma	
• Glioblastoma Multiformi	
• Oligodendroglioma	

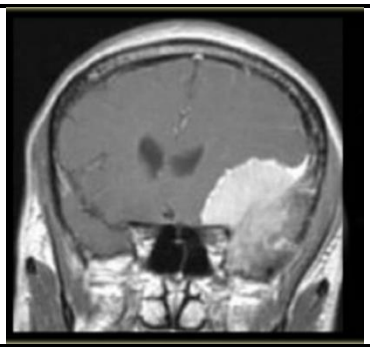
Q2: Location?



❖ Intra-axial Vs Extra-axial Lesions

- CSF cleft (yellow arrow) between the mass and the brain.
- Subarachnoid vessels displaced by the lesion (blue arrow).
- Gray matter between the lesion and the white matter (curved arrow).
- Dural base
- Bone hyperostosis





Q3: Specific CT & MRI features ?

- ❖ **Multiple lesions**
- ❖ **Lesions crossing midline**
- ❖ **Cortical based lesions**
- ❖ **High T1**
- ❖ **Low T2**
- ❖ **Fat**
- ❖ **Calcification**

A) Multifocal Lesions:

- Metastasis
- Lymphomas
- Multicentric glioblastomas and gliomatosis cerebri
- Small vessel disease
- Septic emboli
- Abscesses
- Demyelinating diseases like MS
- Phacomatoses:
  - Neurofibromatosis II
  - Tuberous Sclerosis
  - Von Hippel Lindau

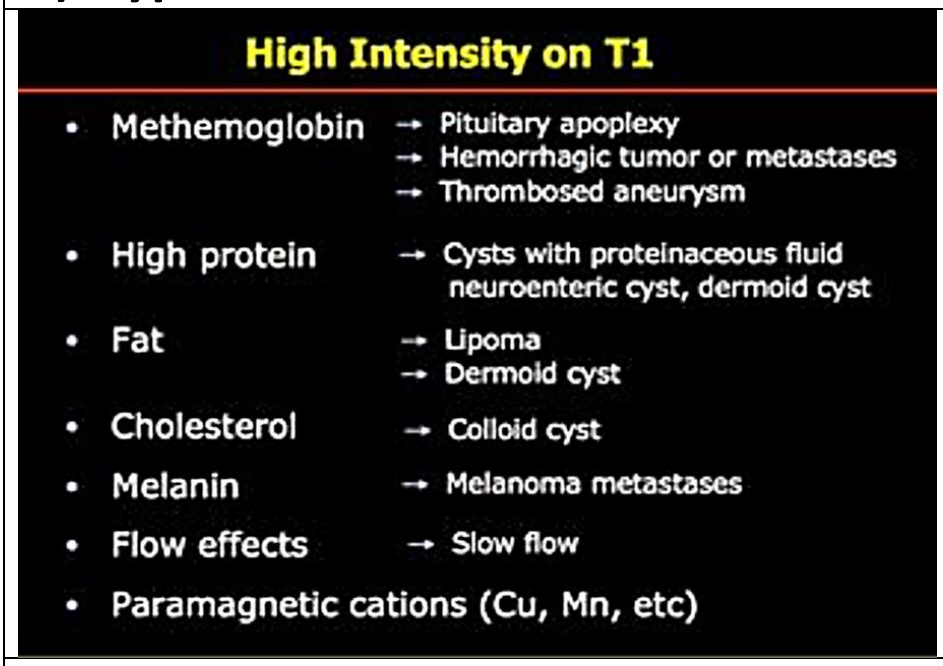
B) Midline Crossing Lesions:

- Glioblastoma multiforme (GBM)
- Radiation necrosis
- Lymphoma
- Tumefactive MS
- Meningioma
- Epidermoid cysts

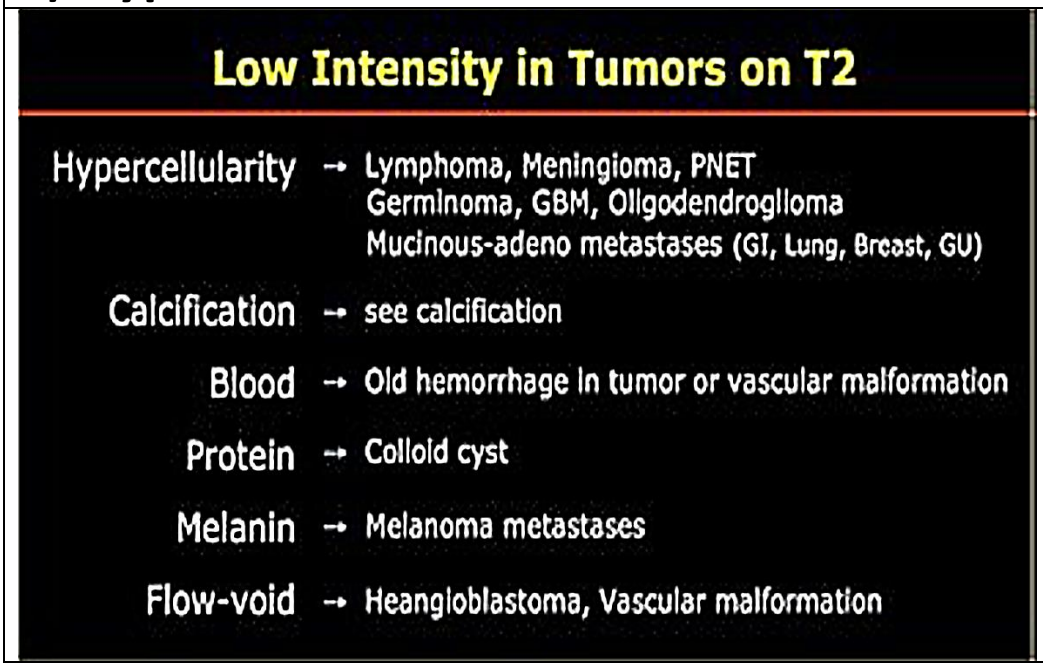
C) Cortical Based Lesions:

- Oligodendroglioma
- Ganglioglioma
- DNET
- PXA

D) Hyperintense T1


High Intensity on T1
• Methemoglobin → Pituitary apoplexy
→ Hemorrhagic tumor or metastases
→ Thrombosed aneurysm
• High protein → Cysts with proteinaceous fluid
neuroenteric cyst, dermoid cyst
• Fat → Lipoma
→ Dermoid cyst
• Cholesterol → Colloid cyst
• Melanin → Melanoma metastases
• Flow effects → Slow flow
• Paramagnetic cations (Cu, Mn, etc)

E) Hypointense T2


Low Intensity in Tumors on T2
Hypercellularity → Lymphoma, Meningioma, PNET
Germinoma, GBM, Oligodendroglioma
Mucinous-adeno metastases (GI, Lung, Breast, GU)
Calcification → see calcification
Blood → Old hemorrhage in tumor or vascular malformation
Protein → Colloid cyst
Melanin → Melanoma metastases
Flow-void → Heangioblastoma, Vascular malformation

F) Calcifications

Intra-axial tumors:

- Astrocytomas (20%)
- Oligodendrogliomas (90%)
- Metastases
- Ependymoma (50%)
- Choroid plexus papilloma (25%)
- Ganglioglioma (40%)

Extra-axial tumors:

- Meningiomas (25%)
- Craniopharyngeomas (90%)
- Chordomas
- Chondrosarcomas

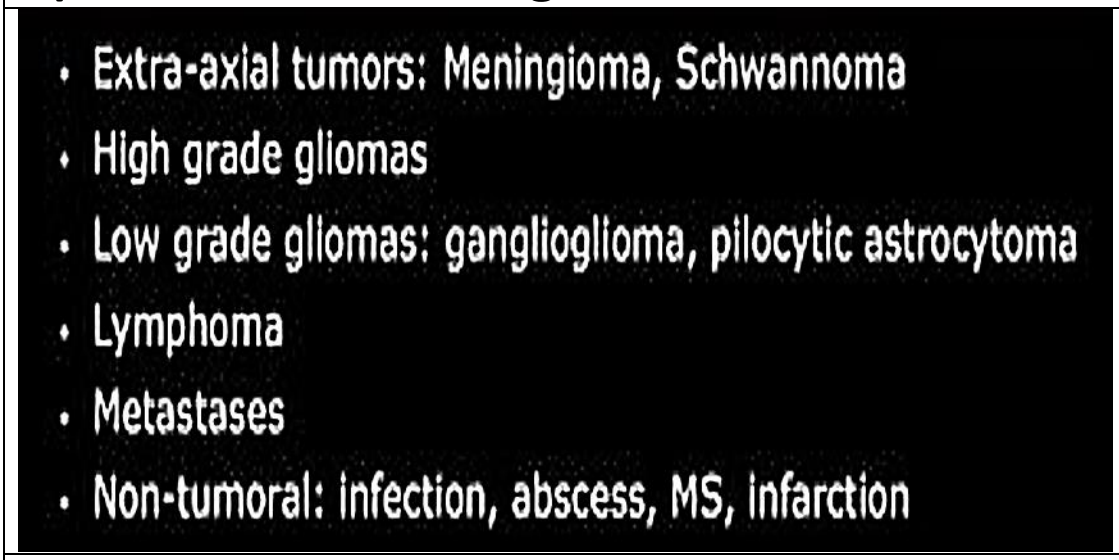
G) Fat

- **Lipoma**
- **Dermoid**
- **Teratoma**

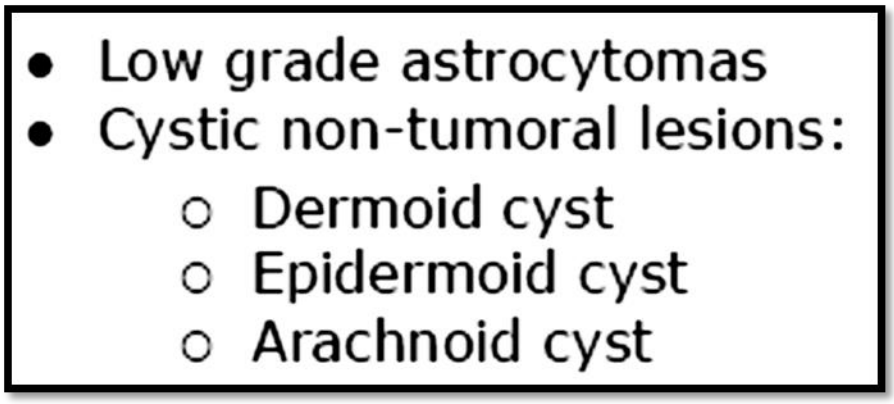
Q4: Contrast Enhanced MRI

- The brain has a unique triple layered blood-brain barrier (BBB) with tight endothelial junctions in order to maintain a consistent internal milieu.
- Contrast will not leak into the brain unless this barrier is damaged

A) Contrast Enhancing Lesions:



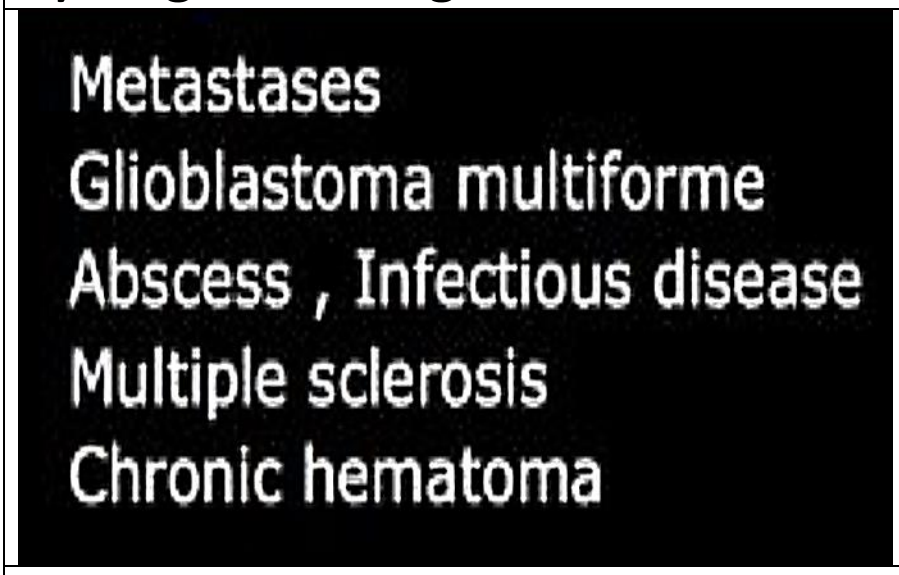
B) Non Enhancing Lesions:



C) Homogeneous Enhancement:

- **Lymphoma**
- **Meningioma**
- **Metastases**
- **Schwannoma**
- **Germinoma**
- **other pineal gland tumors**

D) Ring Enhancing Lesions:



E) Cyst + Enhancing Mural Nodule

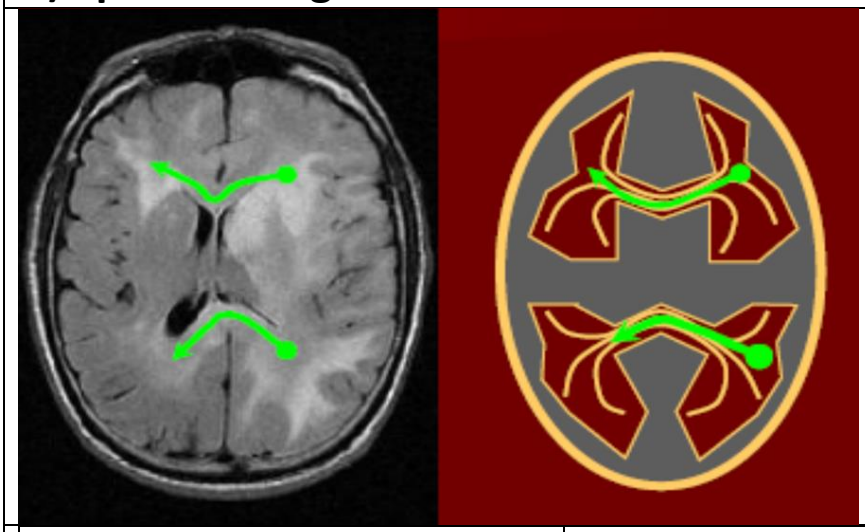
- **Pilocytic Astrocytoma**
- **Hemangioblastoma**
- **PXA**
- **Ganglioglioma**

Q5: Specific Locations

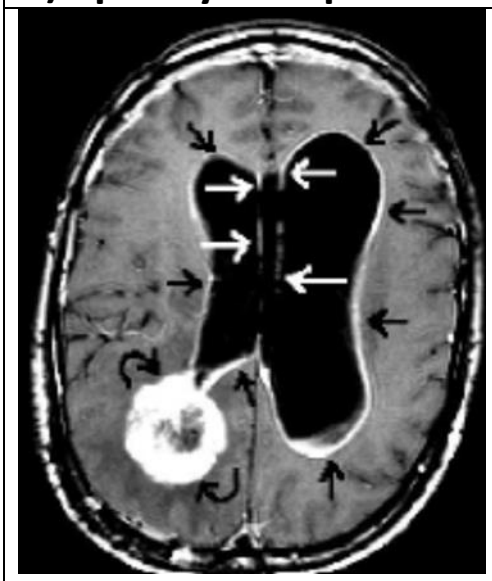
- Sellar – Suprasellar – Parasellar
- Pineal region
- CPA region
- Intraventricular
- Posterior fossa
- Temporal lobe

Q6: Modes of Spread

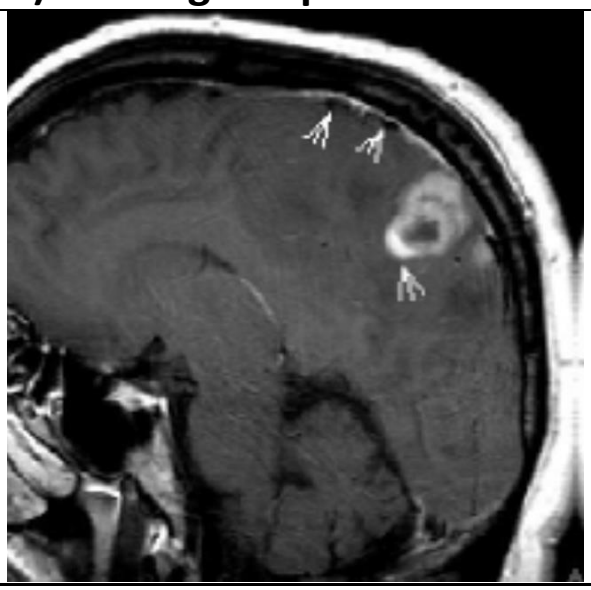
A) Spread along WM tracts



B) Ependymal spread



C) Meningeal spread



Q7: Grading?

WHO Classification:

❖ Astrocytic Tumors

- Grade I: - Pilocytic astrocytoma, pleomorphic xanthoastrocytoma, subependymal giant cell astrocytoma, and subependymoma.
- Grade II: Low-grade fibrillary astrocytoma, mixed oligoastrocytoma
- Grade III: - Anaplastic astrocytoma
- Grade IV: - Glioblastoma multiforme

❖ Oligodendroglial Tumors:

- Oligodendroglioma (WHO grade II)
- Anaplastic oligodendroglioma (WHO grade III)

❖ Ependymal Cell Tumors

- Ependymoma (WHO grade II)
- Anaplastic ependymoma (WHO grade III)
- Myxopapillary ependymoma
- Subependymoma (WHO grade I)

WHO Grading:

- Grade I:
  - Pilocytic astrocytoma (JPA)
  - Pleomorphic xanthoastrocytoma (PXA)
  - Subependymal giant cell astrocytoma (SEGA)
  - Subependymoma.
- Grade II:
  - Low-grade fibrillary astrocytoma, mixed oligoastrocytoma
  - Oligodendroglioma
  - Ependymoma
- Grade III:
  - Anaplastic astrocytoma
  - Anaplastic oligodendroglioma
  - Anaplastic ependymoma
- Grade IV: Glioblastoma multiforme

	High Grade	Low Grade
Conventional MRI	• Mod/ Marked edema • Mod/ Marked mass effect • hge / necrosis	• No / Mild edema • No / Mild mass effect • No hge / necrosis
Contrast MRI		
MRS	• Cho/NAA > 2.2 • Lact/Lipid	• Cho/NAA < 2.2 • Myoinositol
DWI	• ADC < 0.98	• ADC > 0.98
MRP	• rCBV > 1.75	• rCBV < 1.75

Q8: Tumor-related Complications?

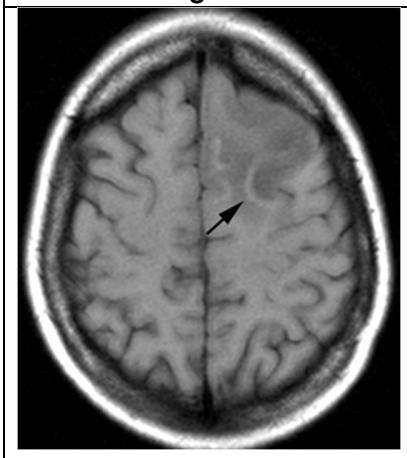
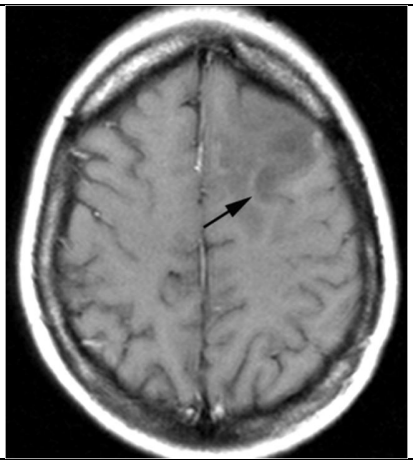
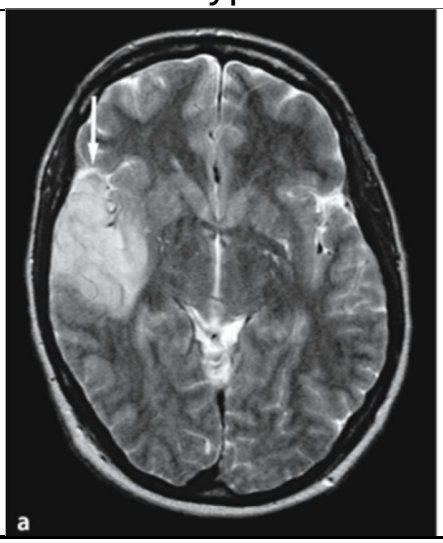
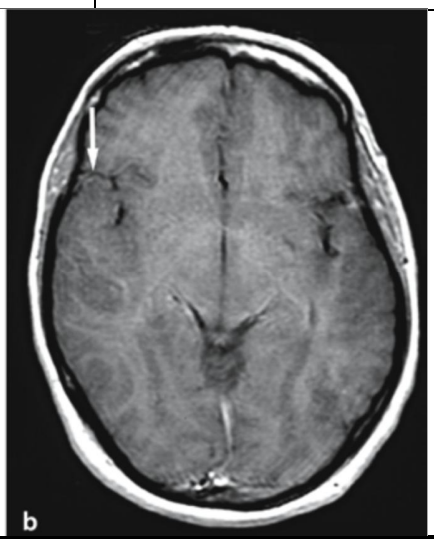
- ❖ **3 emergent complications of brain tumor 3 H's:**
- ➢ **Hemorrhage**
- ➢ **Hydrocephalus**
- ➢ **Herniation**

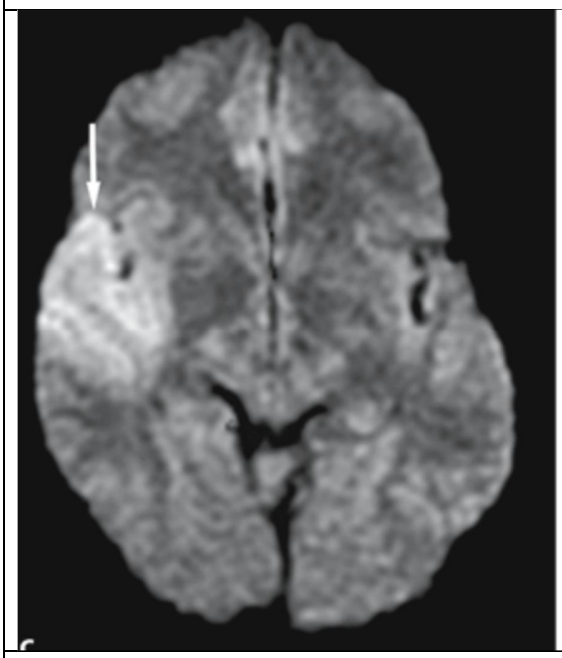
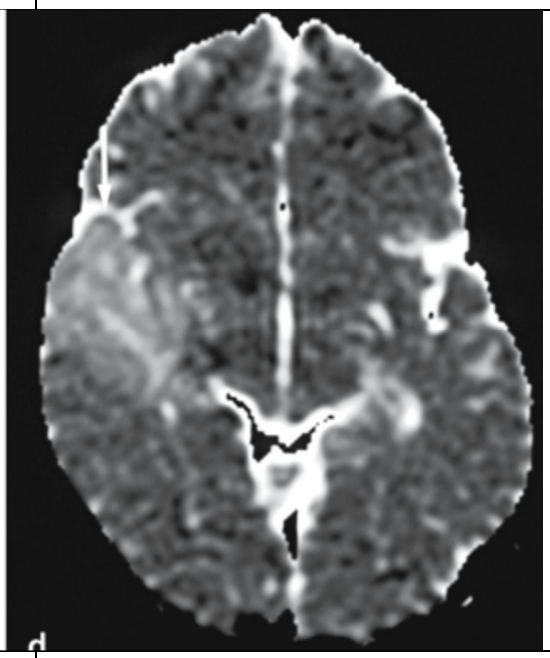
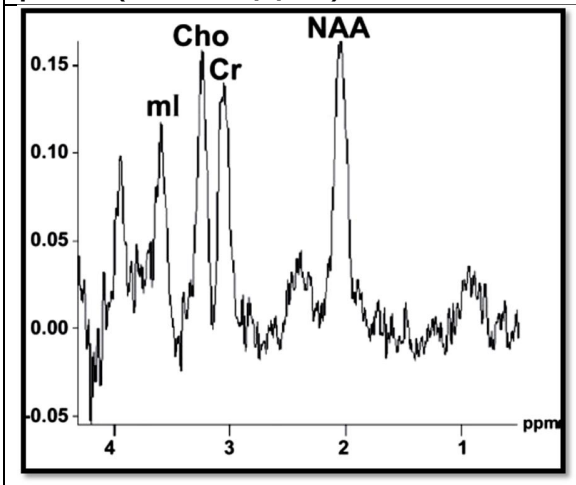
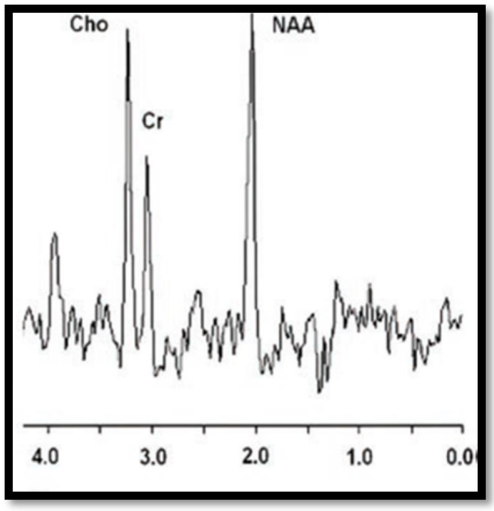
I- Adult Supratentorial Intra-axial Lesions

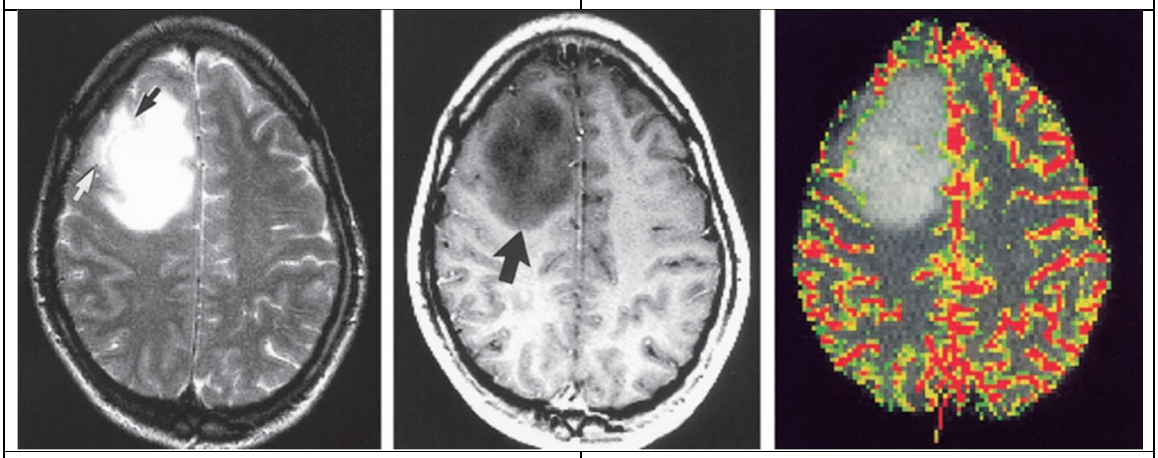
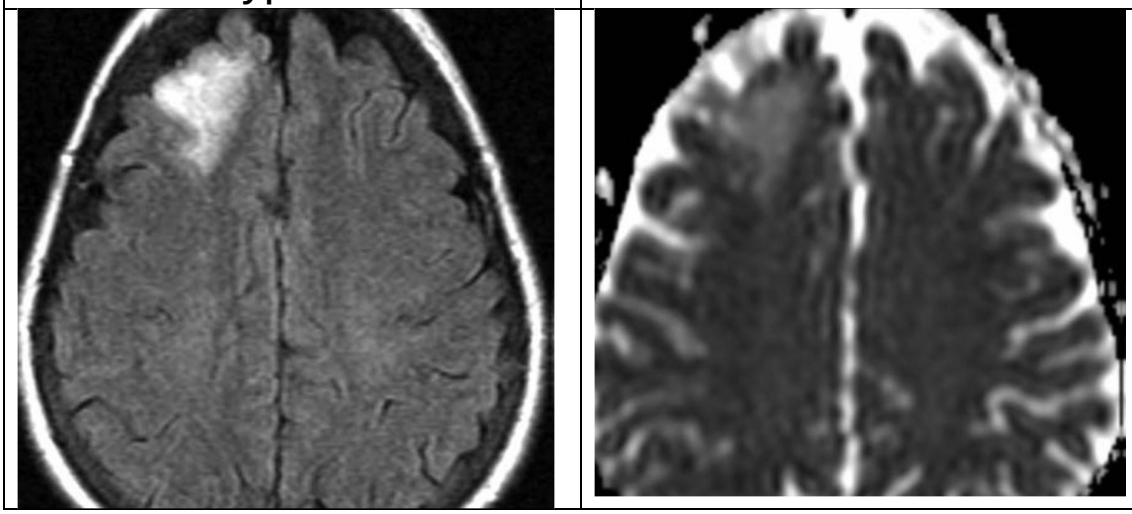
	Low grade Astrocytoma	Anaplastic Astrocytoma	GBM
Grade	Grade II	Grade III	Grade IV
Incidence	20% of astrocytoma	30% astrocytoma	50% astrocytoma
Age	20 – 45 Y	40-60 Y	> 50 Y
Site	Frontal , Temporal	Frontal, Temporal	Frontal, Temporal
	Ill defined	ill defined infiltrative	Ill defined infiltrative
	Homogeneous	Heterogeneous	Heterogeneous
Edema Mass effect Hge Necrosis	No / Mild	Mild / Moderate	Marked
Ca+	Rare		
MRI	T1: Low T2/FLAIR: High		
+C	No/ Mild	No/Mild Heterogeneous	Heterogeneous marginal
ADC	> 0.98		< 0.98
MRS	Ch/NAA < 2.2 High Myoinositol	Ch/NAA >2.2 Low Myoinositol Lipid and lactate peak	
Perfusion	rCBV < 1.75	rCBV > 1.75	
PETCT SUV	Low grade tumor = WM	High grade tumor / WM > 1.5 High grade tumor / GM > 0.6	

1) Low Grade Astrocytoma WHO Grade I

- Diffuse or focal non enhancing hemispheric WM mass
- Age: 20-45 y
- Location: Supratentorial: Frontal & Temporal lobes
- Homogenous ill defined lesion
- No / Mild edema
- No / Mild mass effect
- No / Mild hemorrhage
- No / Mild necrosis
- No / Mild enhancement
- Malignant progression tends to occur after mean time interval of 4-5 y to anaplastic astrocytoma

T1: Low signal	+C: No enhancement
	
T2/FLAIR: Hyperintense	+C: No enhancement
	

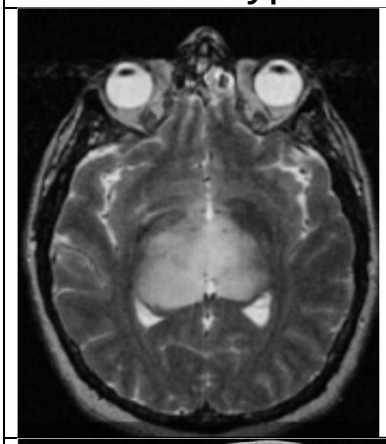
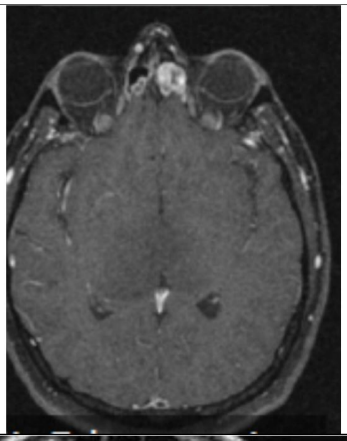
DWI	ADC > 0.98
	
MRS: Elevated myoinositol (ml) peak (at 3.55 ppm)	Cho/NAA < 2.2
	

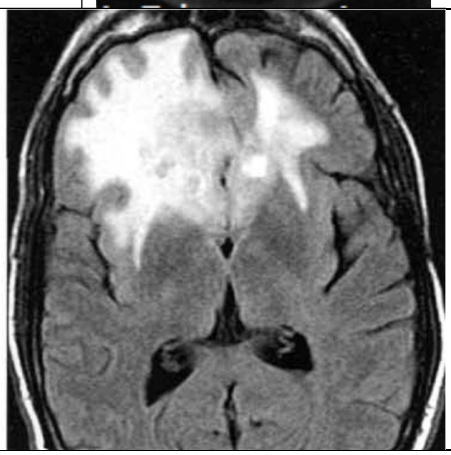
No enhancement	rCBV < 1.75
	
T2/FLAIR: hyperintense	ADC > 0.98
	

2) Anaplastic Astrocytoma WHO Grade III

- ILL defined Infiltrating mass involve WM.
- Age: 40-50 y
- Location: Supratentorial: Frontal & Temporal lobes
- Heterogeneous ill defined infiltrative lesion
- Mild / Moderate edema
- Mild / Moderate mass effect
- Mild / Moderate hemorrhage
- Mild / Moderate necrosis
- No / Mild heterogeneous enhancement
- No restricted diffusion

T2/FLAIR: Hyperintense	+C: No enhancement



3) Glioblastoma Multiforme WHO Grade IV

❖ It is the most common primary CNS malignancy

- Age: > 50 y
- Location: Supratentorial: Frontal & Temporal lobes
- Heterogeneous ill defined infiltrative lesion
- Moderate / Marked edema
- Moderate / Marked mass effect
- Moderate / Marked hemorrhage
- Moderate / marked necrosis

❖ <u>Contrast:</u>

- Peripheral irregular ring enhancement with necrotic core

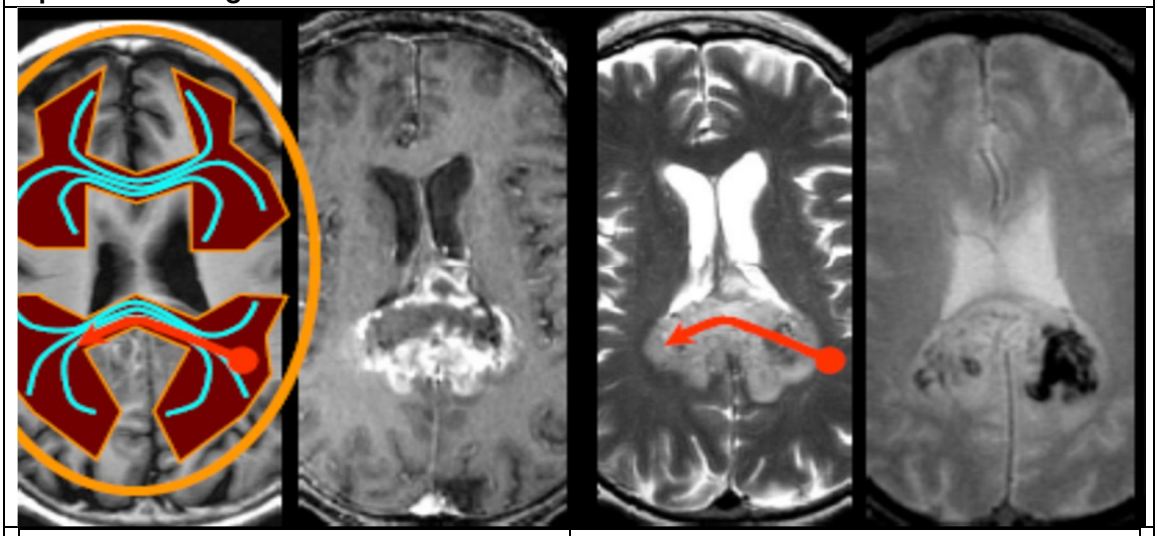
❖ <u>Spread along:</u>

→ Corpus callosum (Butterfly glioma)
→ WM tracts
→ Subependymal seeding
→ Subarachnoid seeding (Leptomeningeal metastasis)

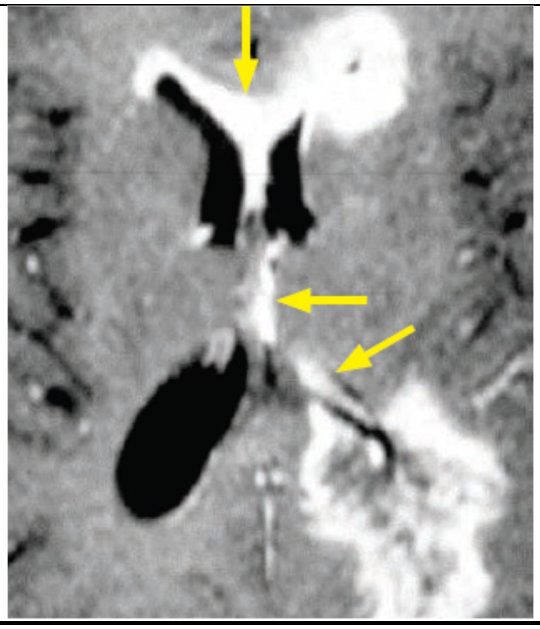
❖ <u>High Grade Glioma:</u>

- MR Diffusion:
  - Solid peripheral enhancing component
    - Diffusion high signal + Low ADC = Restricted
    - ADC < 0.95
  - Necrotic central non-enhancing component
    - Diffusion low signal + high ADC = Facilitated
- MRS:
  - Cho / NAA > 2.2
  - Low myoinositol peak
  - Lipid and Lactate peak
- MR Perfusion:
  - rCBV > 1.75
- PETCT:
  - High grade tumor / WM > 1.5
  - High grade tumor / GM > 0.6

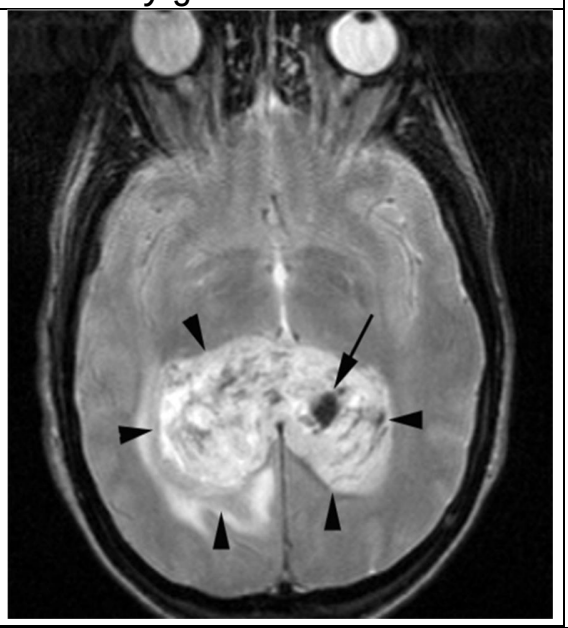
Spread along WM tracts

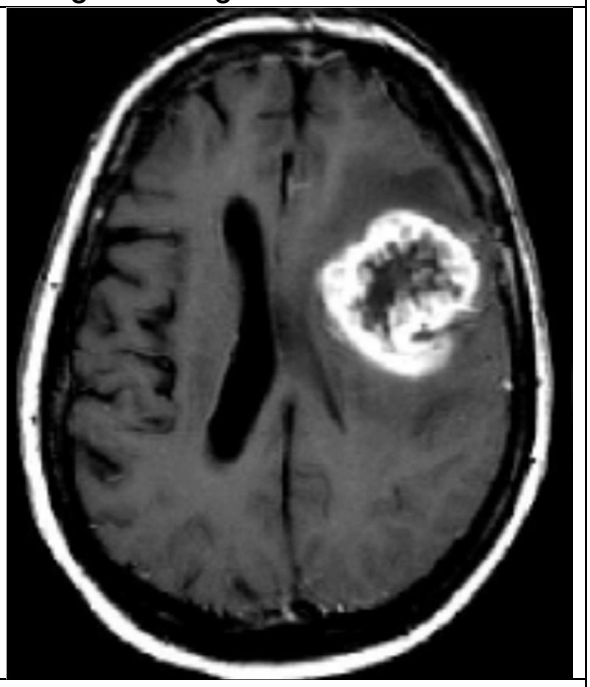
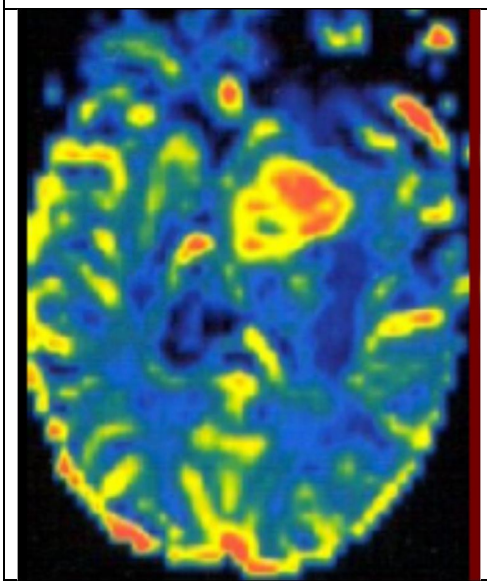
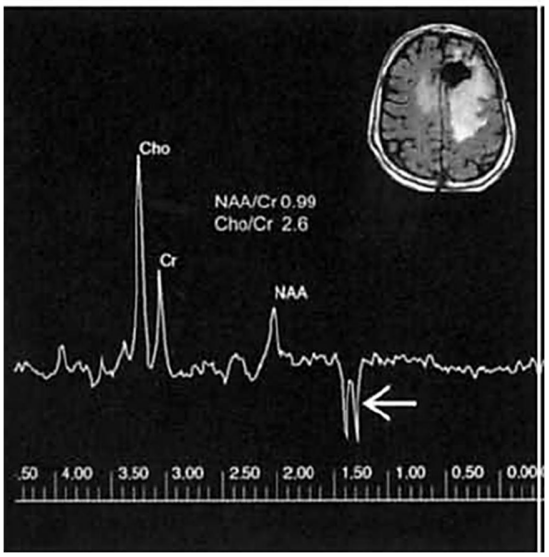


Transependymal spread



Corpus callosum spread = Butterfly glioma



Perifocal edema + Mass effect	Irregular ring enhancement
	
rCBV > 1.75	**MRS**
	

4) Gliomatosis Cerebri Grade III

- Age peak 40-50 Y

- Ill defined infiltrative lesion
- 2 or more lobes
- Bilateral; may cross corpus callosum or massa intermedia
- Loss of GM/WM differentiation.

- T1: Hypointense
- T2: Hyperintense
- Mild mass effect
- NO / minimal enhancement

DWI:

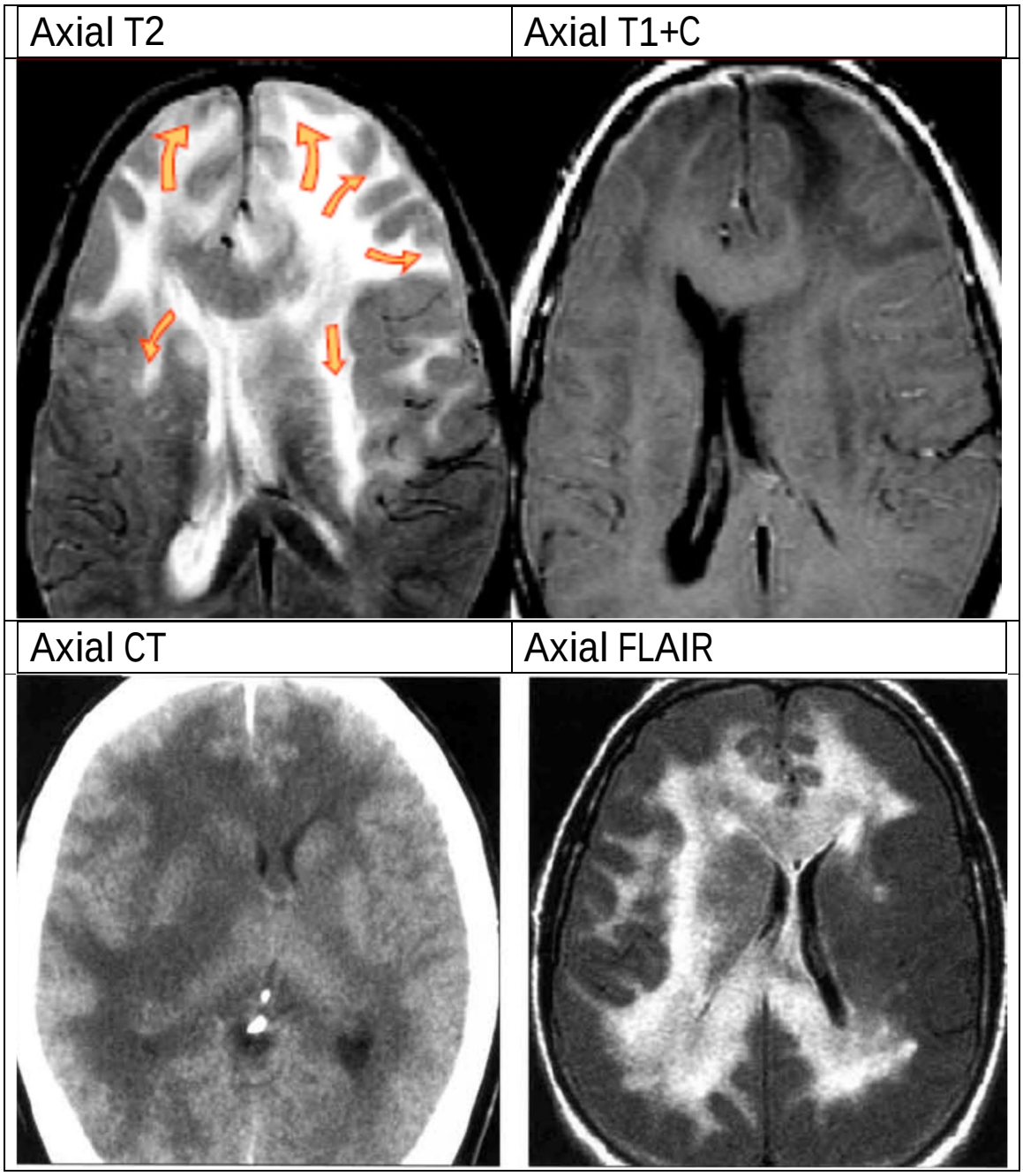
- Usually no restriction

MRS

- Marked elevation of myo-inositol (ml)
- Decreased NAA
- Normal/mildly increased choline (Cho)
- +/- Lactate, lipid peaks at 1.33 ppm

PET:

- FDG shows marked hypometabolism


Axial T2
Axial T1+C
Axial CT
Axial FLAIR

5) Oligodendroglioma

- Well differentiated oligodendroglioma (Grade II)
- Anaplastic oligodendroglioma (Grade III)

- Rare tumor

- Middle age adults

Location:

- Supratentorial cortical and subcortical WM
- Most common site is frontal lobe

- Well demarcated infiltrative mass
- *Calcification: Coarse and dense (70-90%)*
- *Adjacent bone remodeling*
- Cystic degeneration.

- Oligodendroglioma is most likely brain tumor to calcify; however, because astrocytoma is much more common, calcified tumor is still more likely to be astrocytoma.

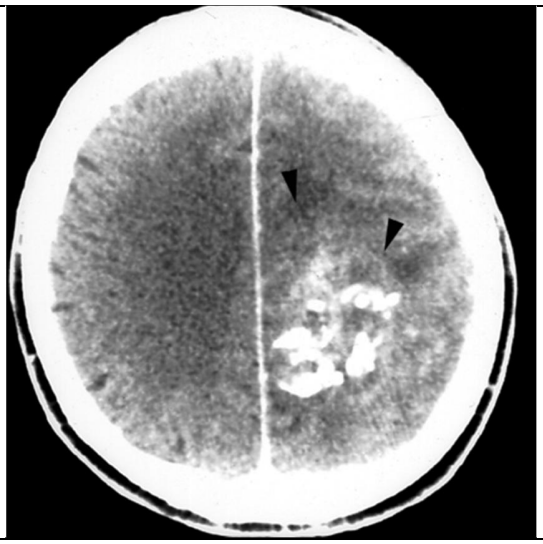
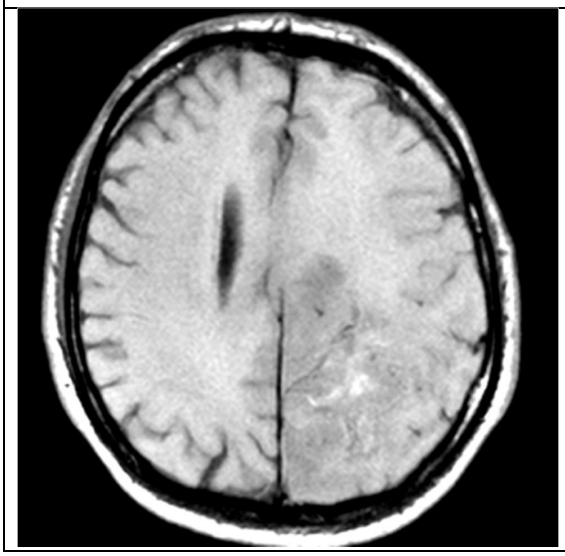
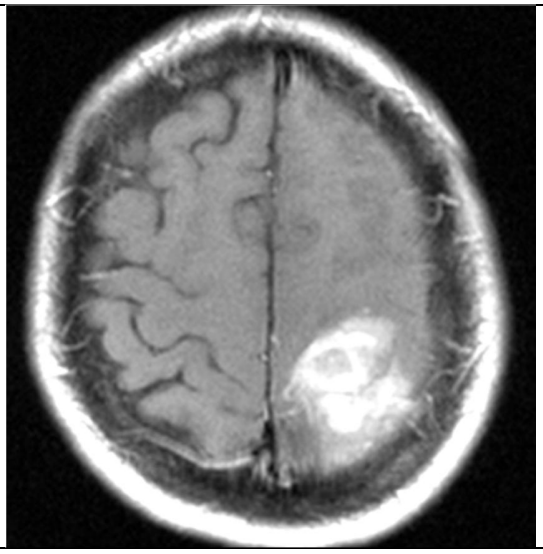
- T1: Heterogeneous Hypointense
- T2: Heterogeneous Hyperintense
- Mild perifocal edema
- Mild mass effect
- +C: Mild / Moderate heterogeneous enhancement

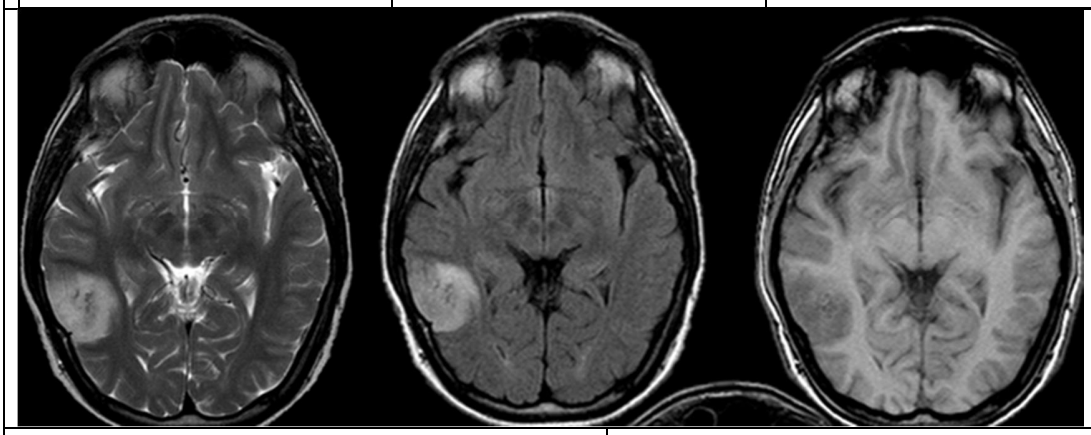
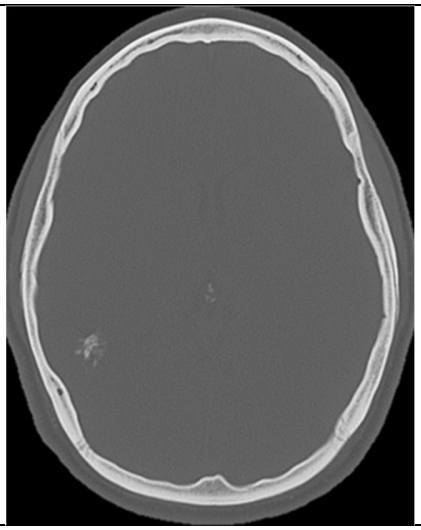
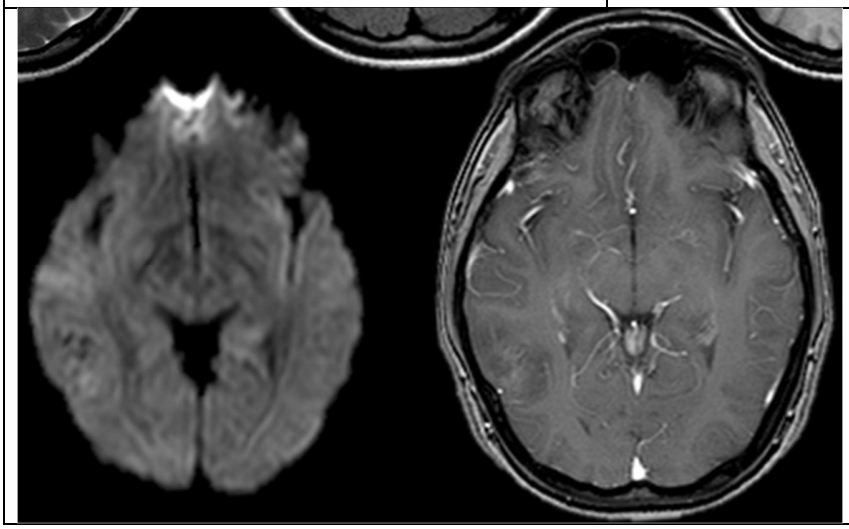
DWI:

- No diffusion restriction

MRS:

- Elevated choline
- Decreased NAA levels

CT Pre Contrast	CT Post Contrast
	
Axial T1	Axial T1 + C
	

Axial T2	Axial FLAIR	Axial T1
		
Axial CT Soft tissue window		Axial CT Bone window
		
DWI: No restriction		+C: Minimal enhancement
		

6) Primary CNS Lymphoma (PCNSL)

- Primary CNS lymphoma (PCNSL) is lymphoma isolated to the CNS
- B-cell non-Hodgkin's type.
- < 2% of malignant primary brain tumors.

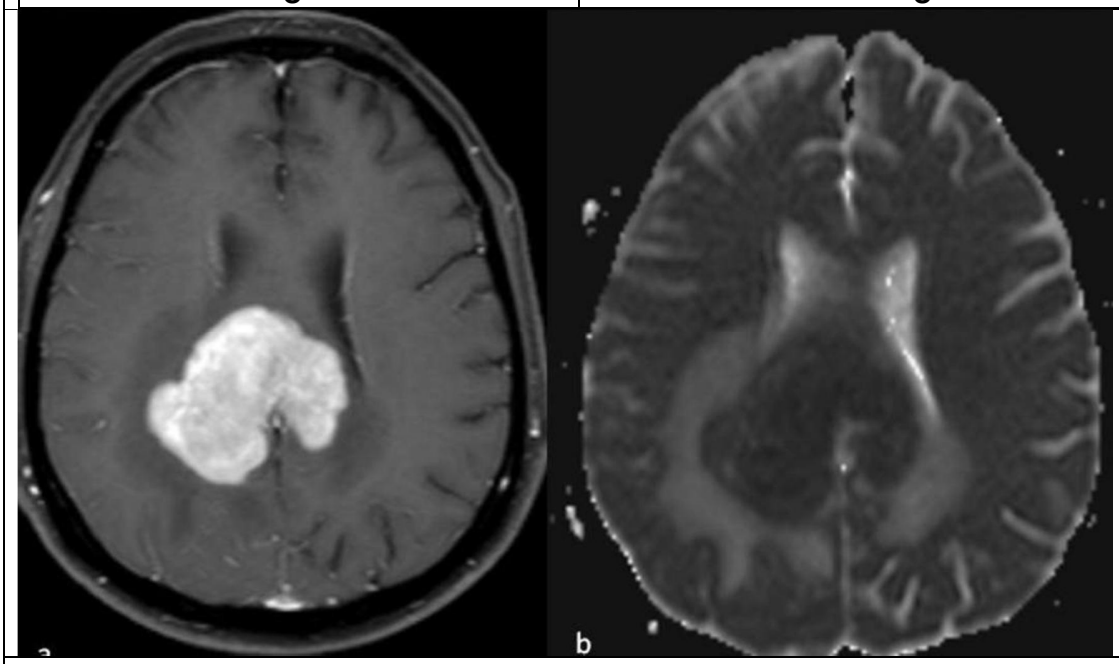
- PCNSL is known to "melt away" with chemoradiation
- The appearance of PCNSL depends on the immune status of the patient.

Primary CNS Lymphoma	Secondary CNS Lymphoma
<u>Parenchymal :</u> • Corpus callosum • Periventricular region • Deep GM structures: • Basal ganglia + Thalami	<u>Leptomeningeal :</u> • Subarachnoid • VR Perivascular • Subependymal infiltration

	PCNSL	High Grade Glioma
Location	Supratentorial: Periventricular / Corpus callosum	
Imaging features	Homogenous No cystic break down No hemorrhage No calcification	Heterogeneous Cystic breakdown Hemorrhage
CT	Hyperdense	Hypodense
T1	Low	Low
T2/FLAIR	Low (High cellularity)	High
+C	Homogenous enhancement (in immunocompetent) Ring enhancement (in immunocompromised AIDS)	Ring enhancement
DWI	Restricted	Peripheral restriction Central facilitated
ADC	Low	Peripheral Low
MRP	rCBV Low <1.8	rCBV High >1.75
MRS	Low NAA, Cr High Cho, Cho/Cr, Cho/NAA ratios Elevated lipid peak	

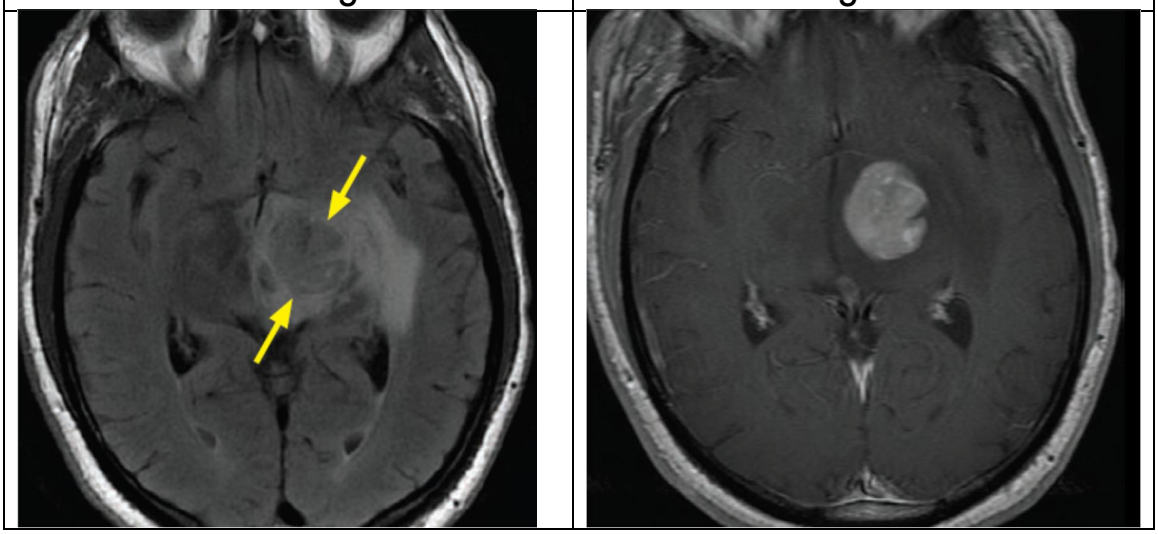
Corpus Callosum PCNSL

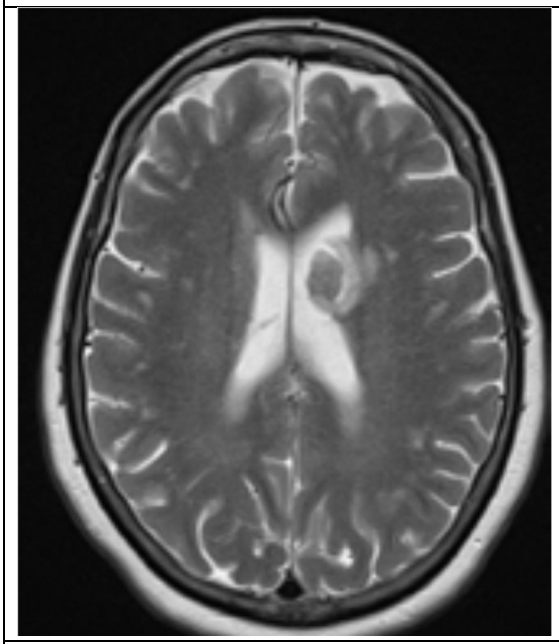
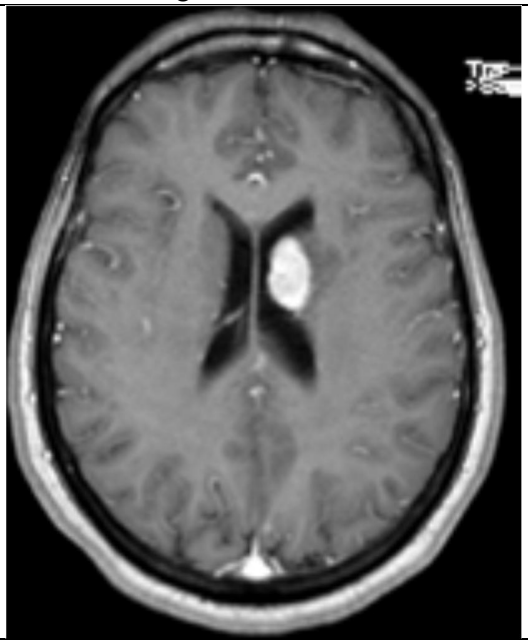
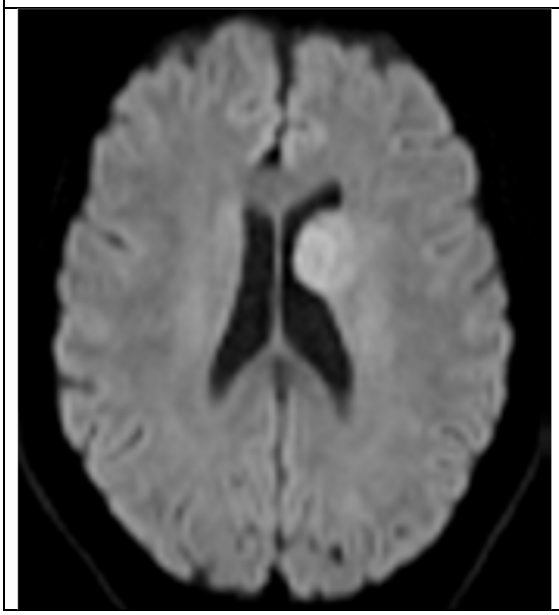
+C: Enhancing lesion	T2/FLAIR: Low signal



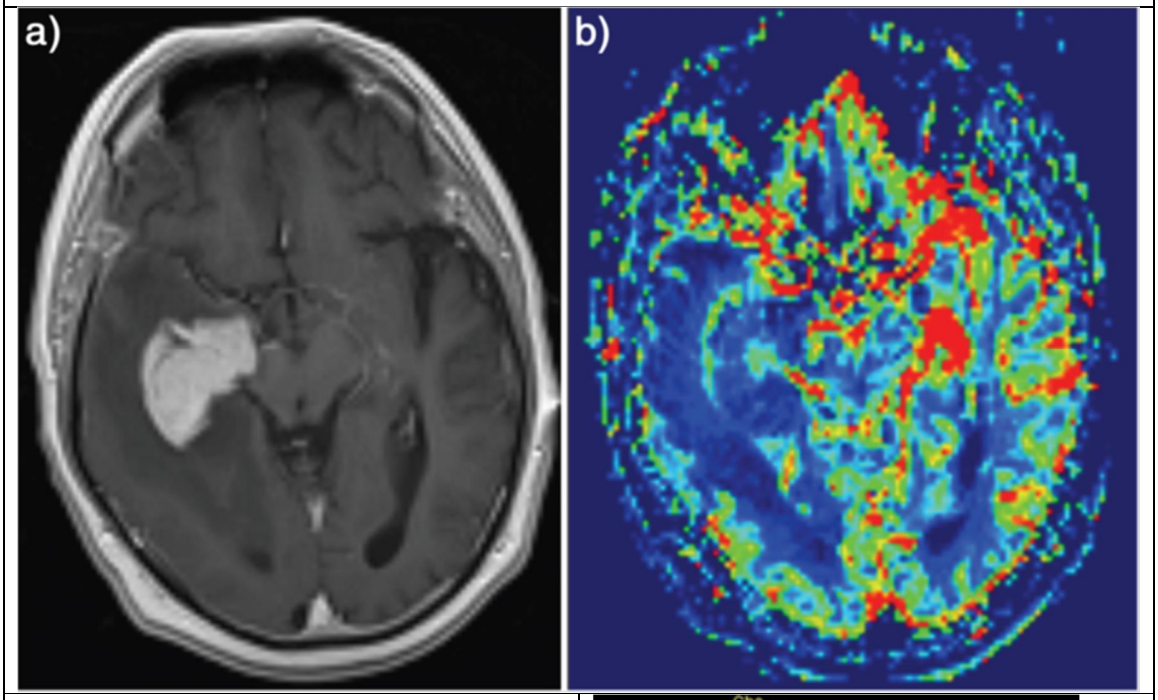
Basal Ganglia PCNSL

T2/FLAIR: Low signal	+C: enhancing lesion



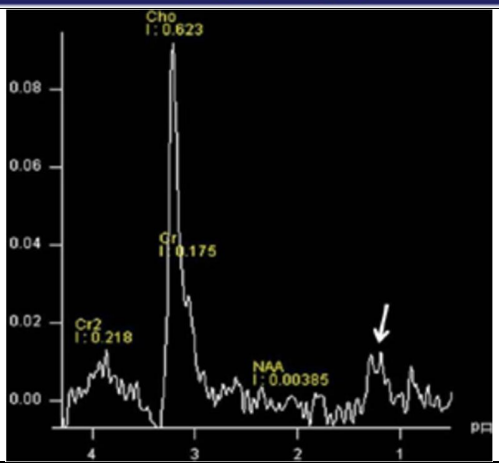
Axial T2: Low	+C: Homogenous enhancement
	
DWI: Restricted	ADC: low
	

MR Perfusion: Low r CBV <1.8



MRS:

- Low NAA
- Low Cr
- High Cho
- High Cho/Cr ratio
- High Cho / NAA ratio
- Lipid peak (Arrow)



7) Intra-Axial Metastasis

- 25-40% of all intracranial neoplasms
- Age > 40 y

Primary :

a) Outside CNS:

- Bronchogenic ca. 50%
- Breast 20% GU 10%
- Melanoma 10% GIT 10%

b) 1ry CNS :

- Medulloblastoma
- Ependymoma
- Choroid plexus papilloma
- Choroid plexus carcinoma
- Germinoma

Location:

a) Parenchymal:
   - Gray-white junction, vascular watershed area, 80% Supratentorial

b) Leptomeningeal.
c) Dural.
d) Skull & vertebrae.

a) Parenchymal Metastasis :

- Multiple 80%
- CT : Variable densities
- T1 : hypo / iso
- T2 : hyper / iso
- Mod / Marked enhancement:-
  - Homogeneous enhancement small lesion
  - Irregular ring enhancement in large lesion
- Marked peripheral oedema

Calcification :-

- Mucinous adenocarcinoma(colon , ovary , pancreas) , osteosarcoma.

Hemorrhagic metastasis:-

- Melanoma, RCC, choriocarcinoma

b) Leptomeningeal = Meningeal carcinomatosis :

- Contrast MRI > CT
- Enhancing basilar cisterns or sulci : typical
- Focal subarachnoid masses : less common
- Carcinomatous encephalitis : rare

c) Dural :

- Contrast MRI > CT
- Diffuse thickening or nodular enhancement of the dura.

d) Skull & Vertebrae :

- CT: - Destructive lytic lesion.
- T1: Hypointense patchy replacement of fatty B.M
- T1+C FS: abnormal enhancement of BM infiltration
- May be associated with epidural mass.
- Dural thickening.

II- Pediatric Supratentorial Lesions

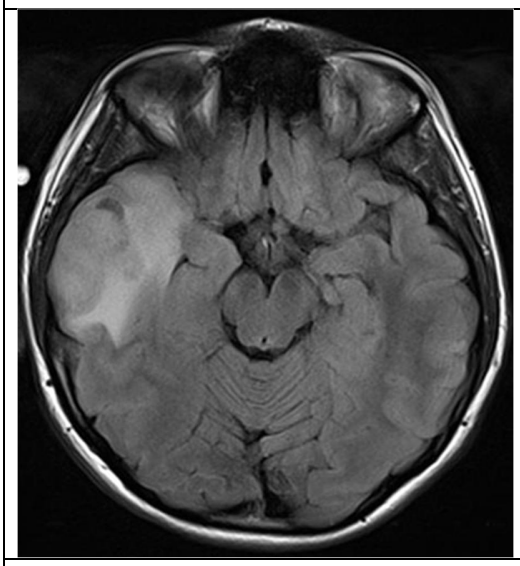
<u>Low Grade Tumors:</u> • Pleomorphic Xanthoastrocytoma (PXA) • Dysembryoplastic neuroepithelial tumor (DNET) • Desmoplastic Infantile Ganglioglioma (DIG) • Ganglioglioma /Gangliocytoma
<u>High Grade Tumors:</u> • Ependymoma • Primitive Neuroectodermal Tumor (PNET) • Atypical teratoid/rhabdoid tumors (ATRT)

	Pleomorphic Xanthoastrocytoma (P X A)	Ganglioglioma Gangliocytoma DIG	DNET
WHO Grade	Low Grade		
Age	Children – young adults		
Site	Cortical + adjacent leptomeningeal Temporal > Parietal, Frontal		
	Temporal lobe Epilepsy		
Rad.	1) Well defined 2) Homogenous 3) Partialy solid / cystic 4) Mural nodule may calcify 5) No hge		1) Well defined 2) Homogenous 3) No cystic 4) No calcify 5) No hge
	No / Minimal perifocal edema No / Minimal mass effect		
CT	Hypodense mural nodule		Hypodense mass
MRI	Solid : - T1 : hypo - T2 : hyper Cyst : CSF like signal		T1 : Hypo T2 : Hyper (Bubbly) FLAIR: Ring sign
+C	Enhancement of mural nodule ± cyst wall		NO / Minimal enhancement

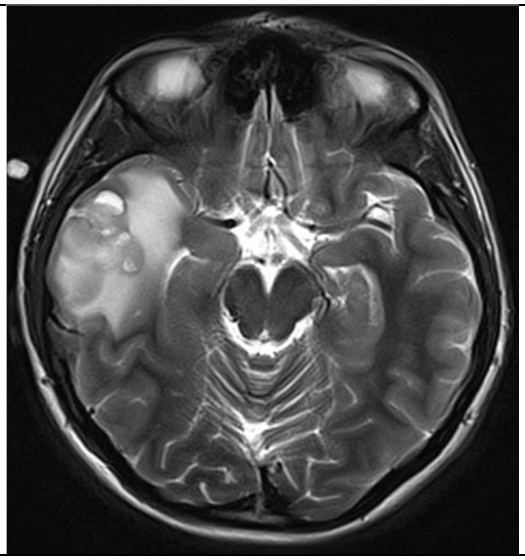
1) Pleomorphic Xanthoastrocytoma (PXA)

- Low Grade (WHO Grade II)
- Location: most commonly in the temporal lobe.
- Well-defined peripheral or cortical lesion
- Partially cystic / partially solid
- Cystic lesion + Enhancing mural nodule
- Adjacent bone remodeling
- Meningeal enhancement
- No diffusion restriction

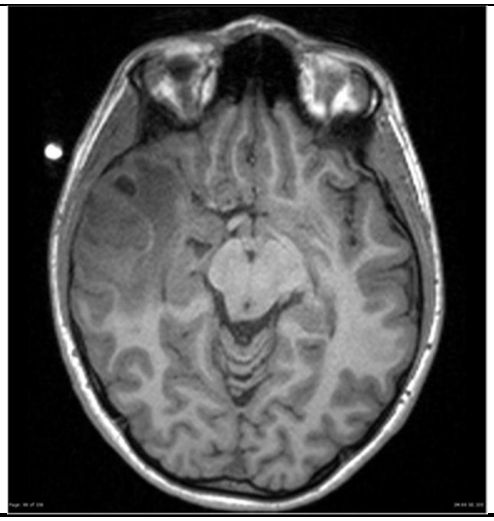
Axial FLAIR



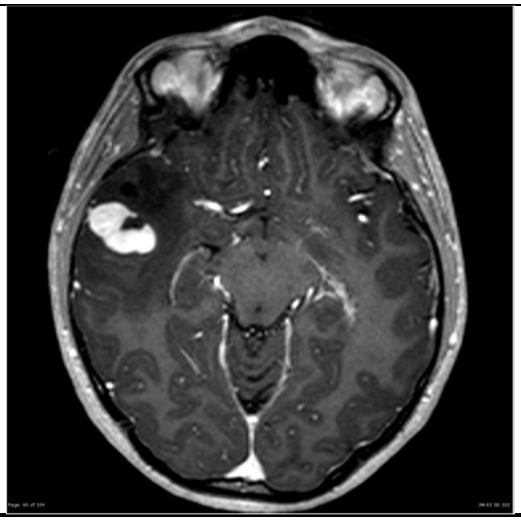
Axial T2



Axial T1



Axial T1+C



2) Dysembryoplastic Neuroepithelial Tumor (DNET)

- WHO Grade I

Location:

- 60% temporal lobe
- 30% frontal lobe
- Cortical and subcortical

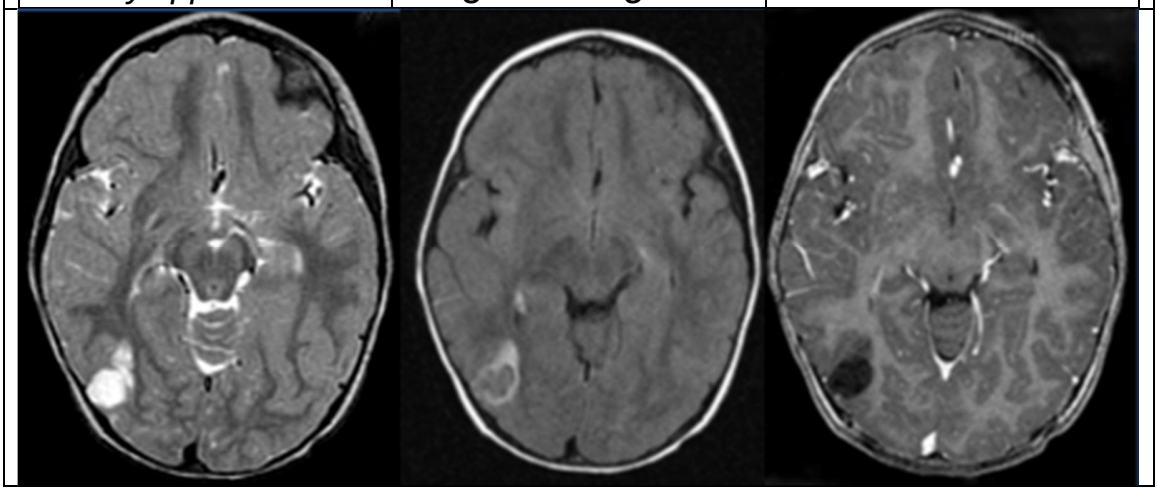
- Cause of 20% cases of medically refractory epilepsy

- Cortical and subcortical wedge shaped lesion
- T1 : Low signal
- T2 : high signal *'bubbly appearance'*
- FLAIR: mixed signal *'bright rim sign'*

- ADC: highest ADC value
- Perfusion: Low rCBV
- Scalloping of inner table skull

- No restricted Diffusion
- No enhancement.
- No vasogenic edema.
- No mass effect

T2 : high signal *'bubbly appearance'*	FLAIR: mixed signal *'bright rim sign.*	No enhancement.

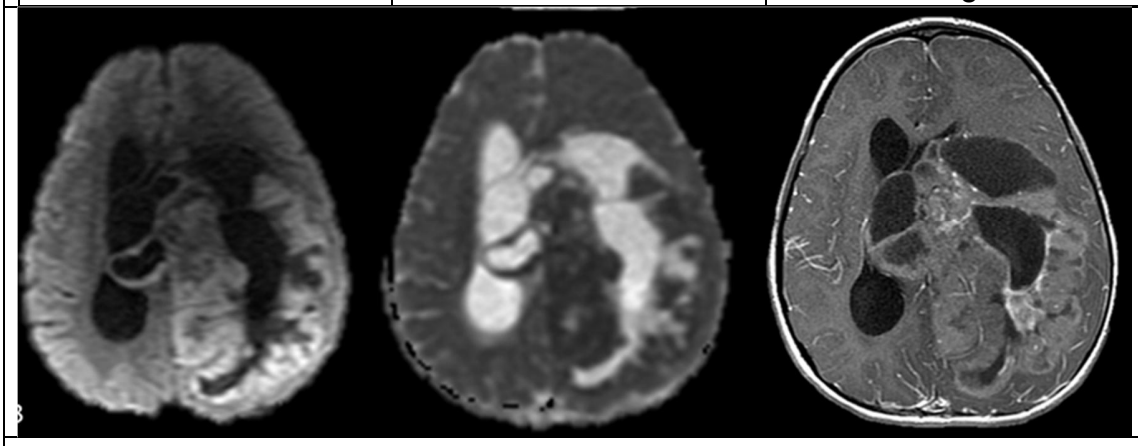


	PNET	AT/RT	Ependymoma
WHO Grade	High Grade (Grade IV)		
Age	< 5 y		
Location	90%: Posterior fossa 10%: Supratentorial extraventricular		
	Calcification / Necrosis / Hemorrhage		
MRI	T1 : Heterogeneous mixed hypo /iso / Hyper signal T2: Heterogeneous high signal FLAIR: Heterogeneous high signal		
+C	Marked heterogeneous enhancement		
	CSF drop metastasis		
DWI	Soli component: Restricted diffusion Cystic component : Facilitated diffusion		
ADC	Solid component: Low ADC value Cystic component : High ADC value		
Perfusion	Increased rCBV		
MRS	High choline Low creatine Low NAA		

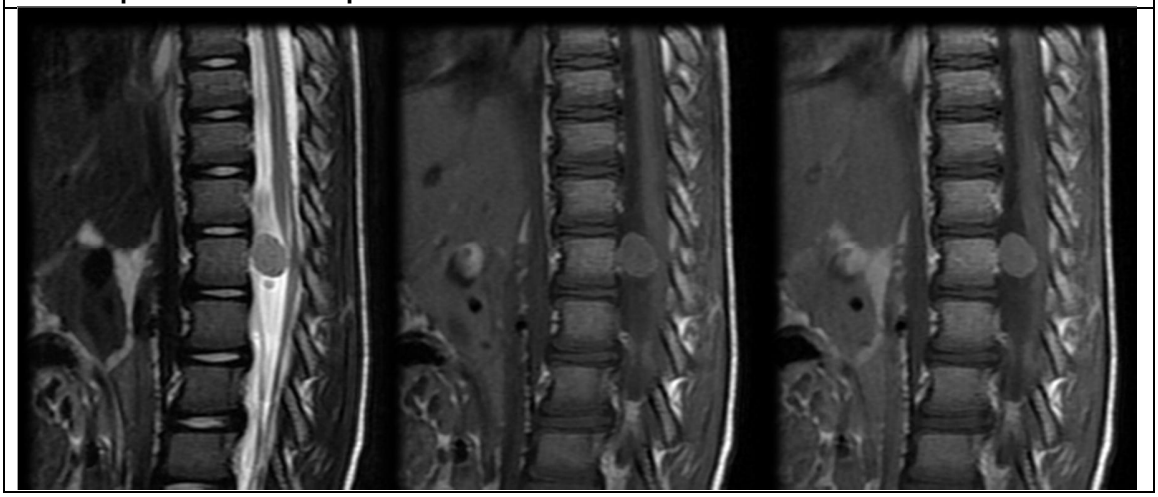
1) Supratentorial PNET

T2: heterogeneous high signal	FLAIR: heterogeneous high signal	T1: heterogeneous high signal

DWI: Restricted	ADC: low value	+C: heterogeneous



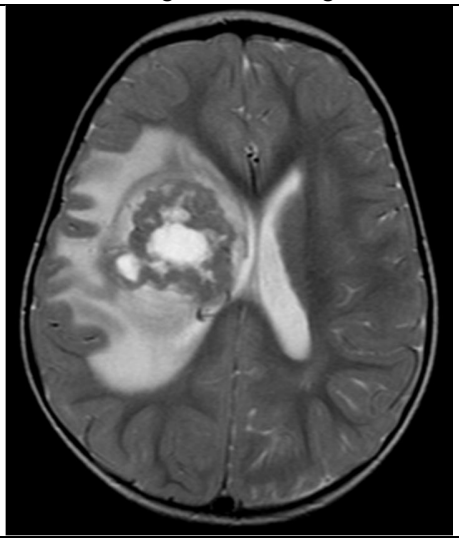
MRI Spine: CSF drop metastasis



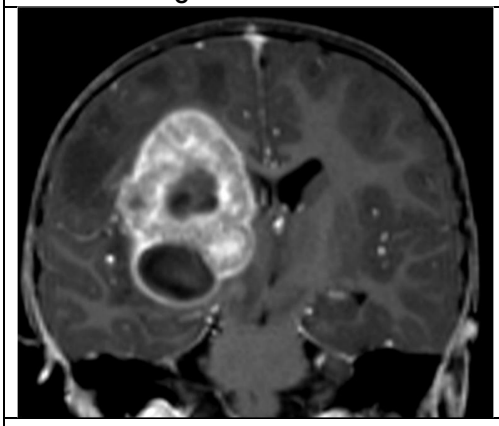
2) ATRT

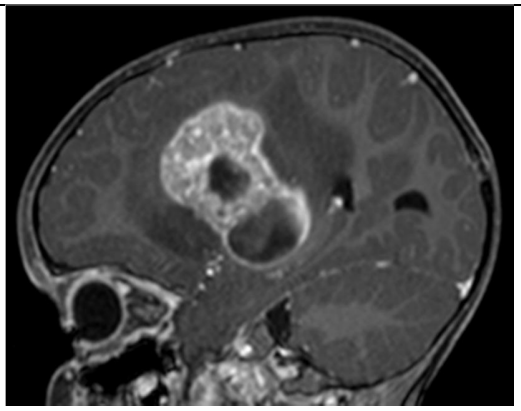
T1: heterogeneous low

T2: heterogeneous high

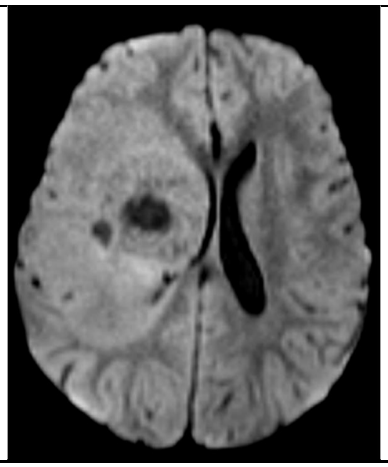


+C: heterogeneous enhancment

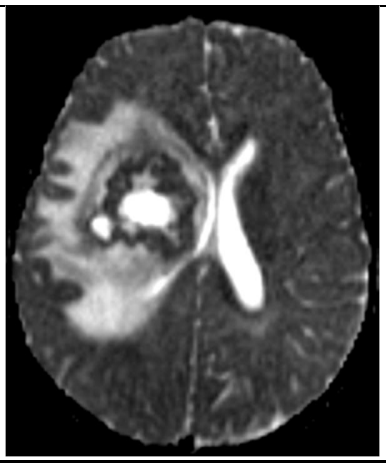




DWI: Restricted

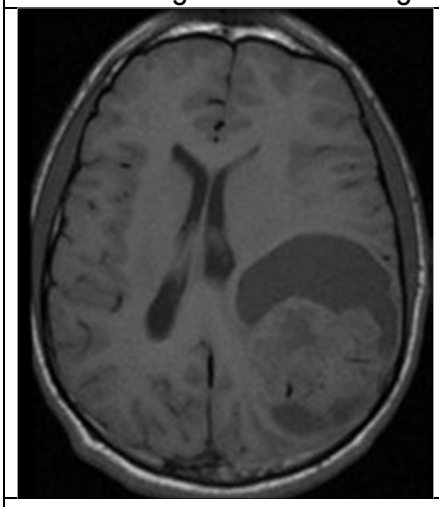


ADC: Low ADC



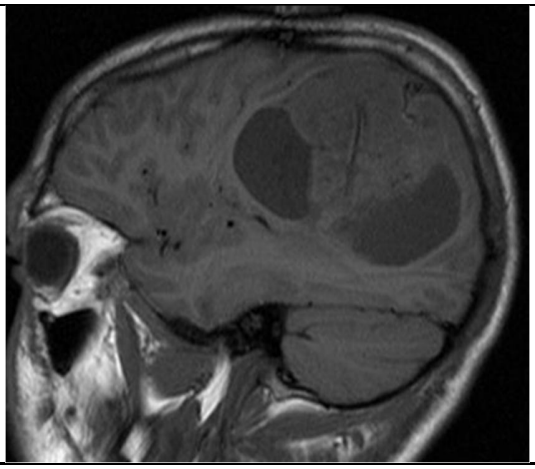
3) Supratentorial Ependymoma

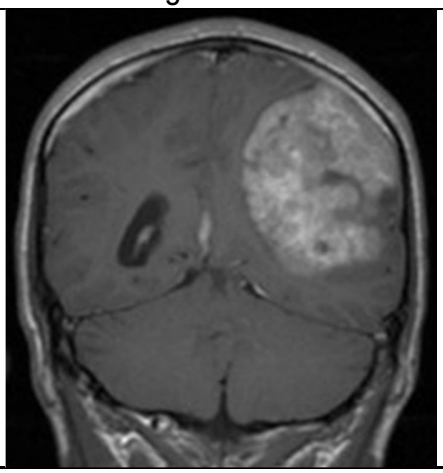
T1: heterogeneous Low signal



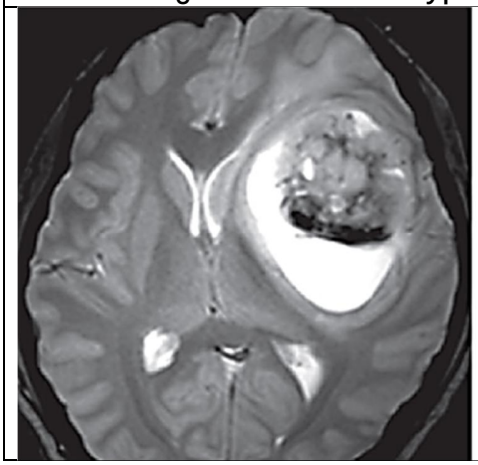
T2: heterogeneous high signal

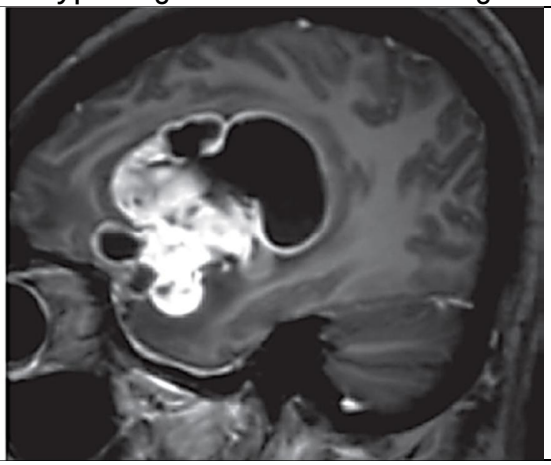
+C: Heterogeneous enh





T2: Heterogeneous mixed hypo + Hyper signal due to hemorrhage





III- Adult Posterior Fossa

❖ Cerebellar Lesions:

1) Metastasis
2) Hemangioblastoma

❖ 4th Intraventricular Lesions:

3) Subependymoma (Solid nonenhancing)
4) Choroid plexus papilloma (solid enhancing)
5) Rosette-forming Glioneuronal Tumor (RGNT) (Cystic)

❖ CPA Lesions (See petrous)

6) Schwannoma
7) Meningioma
8) Epidermoid
9) Arachnoid
10) Endolymphatic sac tumor / VHL
11) Vascular loop / aneurysm

❖ Hemangioblastoma

Incid : Most common 1ry cerebellar tumor in adults.

Age : Middle age adults (30 – 65 y)

Association : *VHL*

Location :

1) Cerebellar hemisphere
2) Spinal cord
3) Medulla

Radiological features: 3 patterns :

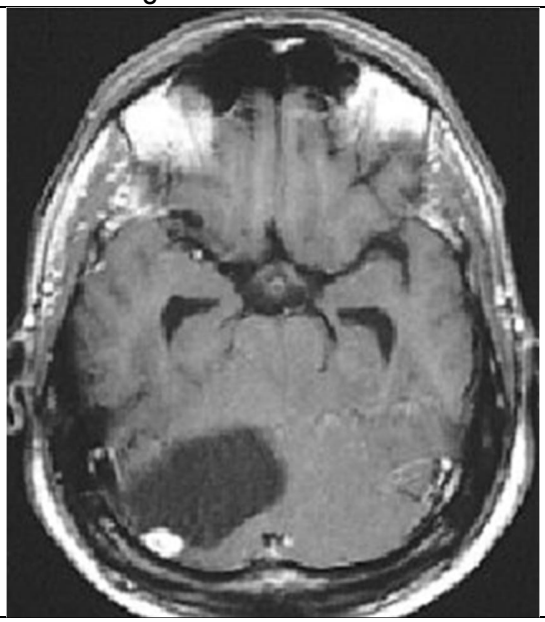
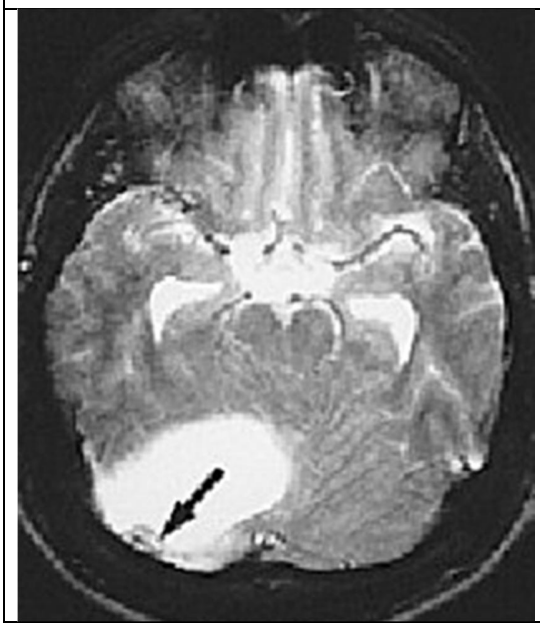
1) Cyst with enhancing mural nodule (75%)
2) Solid enhancing neoplasm (10%)
3) Enhancing lesion with multiple cystic areas (15%)

MRI :

- Solid component:
  - T1 : Iso
  - T2 : Hyper
- Cystic component:
  - CSF like signal

+C:

- Mural nodule :
  - Strong enhancement
- Cystic component:
  - No enhancement
  - Mural enhancement

Axial T1	Axial T1+C: Cystic lesions + Enhancing mural nodule
	
Axial T2	Angiography
	

IV- Pediatric Posterior Fossa Tumors

- Medulloblastoma (PNET)
- Ependymoma
- Juvenile Pilocytic Astrocytoma (JPA)
- Brain stem glioma
- Tectal glioma
- AT/RT

	Medulloblastoma	**Ependymoma**
Incid	**1st common**	**4th common**
WHO	**Grade IV**	**Grade II**
Age	**2 peaks :** 75% Children < 15 Y 15% Adults 25-30 Y	**2 peaks** : Children 1-5 Y Adult mid 30 Y
Location	**90% Posterior fossa** **10% Supratentorial**	**60% Posterior fossa** **40% Supratentorial**
Site	1) Roof 4th ventricle 2) Cerebellar vermis 3) Caudal brain stem 4) Cerebellar hemisphere	1) Floor 4th ventricle 2) F.luschka 3) CPA
Rad	- Well defined - ***Homogeneous*** - Cystic areas - No calcification - No hge	- Well defined - ***Heterogeneous*** - ***Cyst formation 50%*** **- Punctuate calcif.50%** - No hge
CT	***Dense cell packing*** ***Hyperdense***	***Hypodense***
MRI	***Dense cell packing*** T1 : hypo ***T2 : Iso***	*** Solid :-** T1 : Hypo T2 : Hyper *** Cyst : CSF signal**
+C	***Intense homogenous*** enhancement	***Mod. Heterogeneous*** enhancement
Mass effect	4th ventricle displaced => Obstructive hydrocephalus	
Spread	a) ***CNS dissemination = CSF seedling*** b) ***Non CNS metastasis*** : Bone metastasis (Sclerotic) Abdominal cavity (via VP shunt)	***Plastic ependymoma*** Extend through 1) F. magnedi => C.Magna 2) F. luschka => CPA
ADC	**< 0.9 X10^{-3} mm^2/s**	**1.0 – 1.3 X10^{-3} mm^2/s**

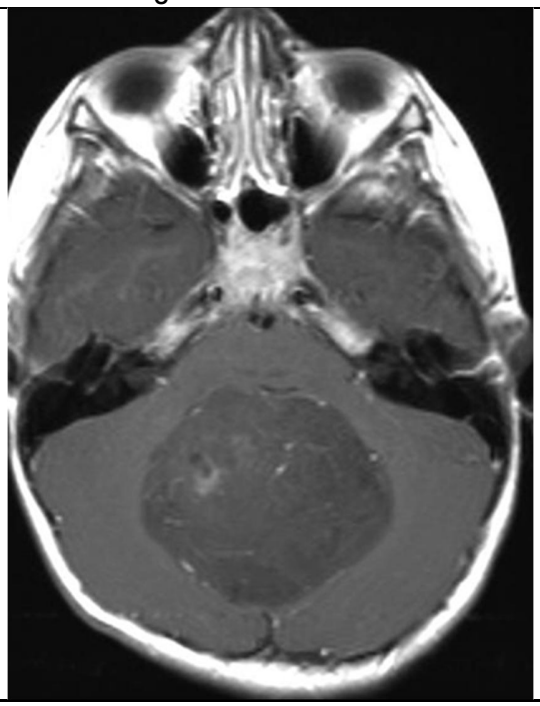
1) Medulloblastoma

- Most common pediatric posterior fossa tumour
- CNS primitive neuroectodermal tumours (PNET)
- WHO grade IV

- Age = 4 – 11 y
- Sex= M>F

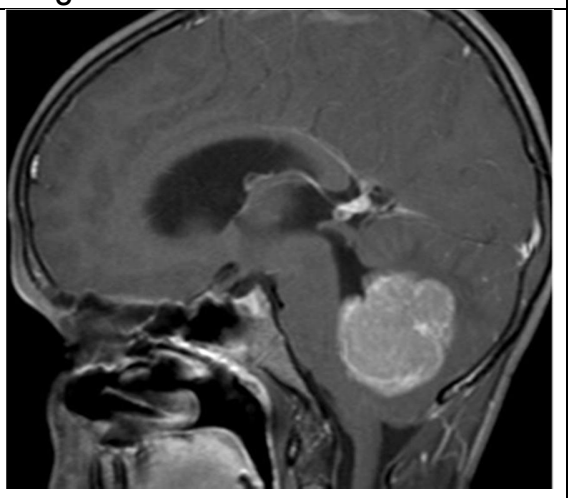
- Midline
- Intraventricular (4th ventricle)
- 2ry hydrocephalus
- Dense cell backing
- Hyperdense
- Homogenous
- 20% Calcification
- Rare cystic changes
- Rare hemorrhagic

T1: Low signal



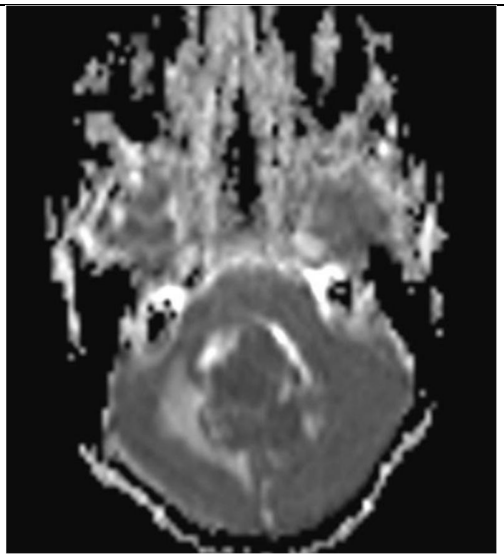
T2: Homogenous Iso – high signal

+C: Homogenous rather than heterogeneous enhancement



DWI: Restricted

ADC < 0.9



2) Atypical Teratoid/Rhabdoid Tumor (ATRT)

- WHO IV
- Aggressive tumor that may appear similar to medulloblastoma, but occurs in slightly younger patients.
- Location: Posterior fossa.
- ATRT is associated with malignant rhabdoid tumor of the kidney

3) Ependymoma

- 3rd most common brain neoplasm in children.
- 3%–5% of intracranial neoplasms.
- 2 age peaks: 1-5yrs and ~35yrs

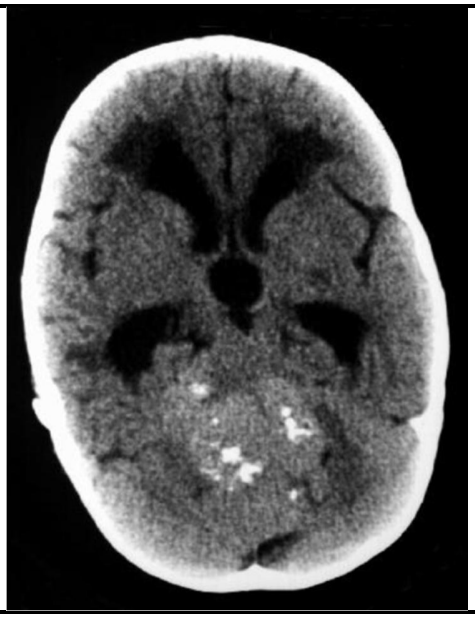
WHO grade

- Most ependymoma are WHO II.
- Myxopapillary ependymomas (thecal sac) are WHO I.
- Anaplastic ependymomas are WHO III.

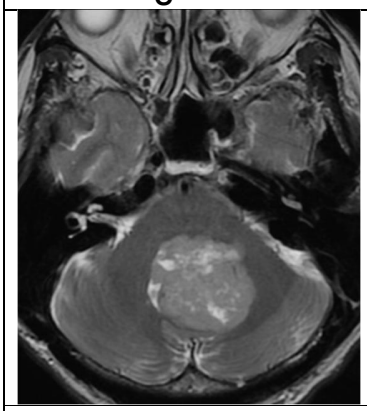
Location:

- 40% Supratentorially.
- 60% Posterior fossa.
  - 75% 4th ventricle
  - 15% CPA
  - 10% Cerebellar

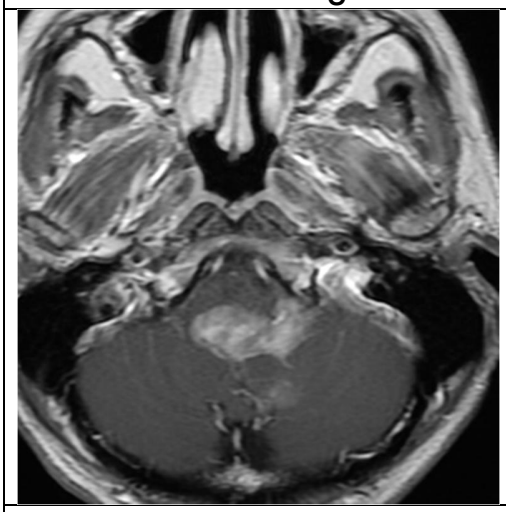
- Heterogeneous
- Cystic components
- Areas of small chunky calcification.
- Occasionally, intratumoral hemorrhage
- hypo-isoattenuating

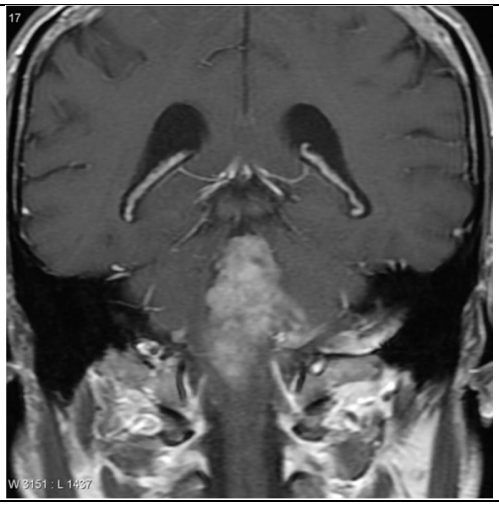


Heterogeneous T2 signal

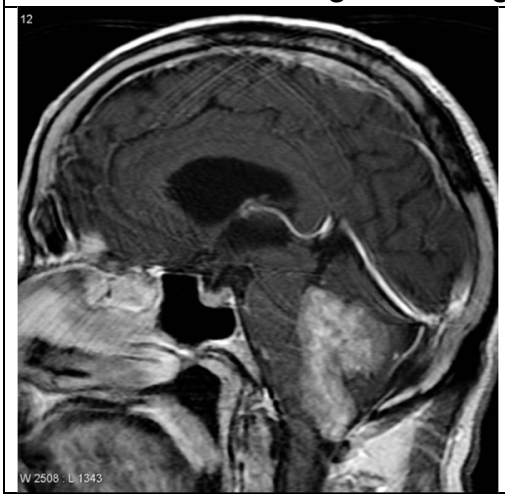


Extension through F. Luschka





Extension through F. magendi



4) Juvenile Pilocytic Astrocytoma (JPA)

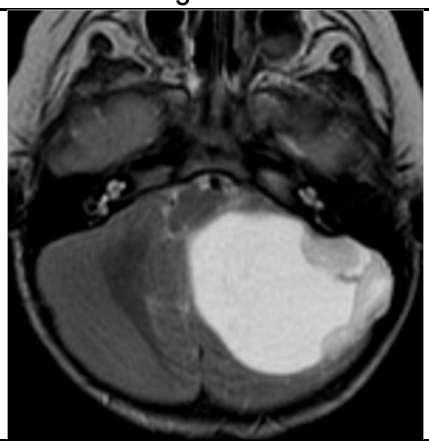
- WHO Grade I
- 2nd common pediatric posterior fossa tumor

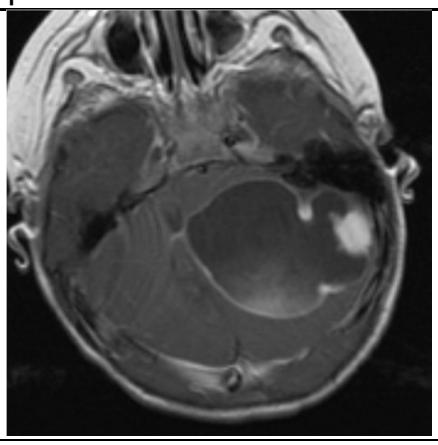
- M=F
- Age = 5-15yrs.

- 40-50% of intracranial neoplasms
- 60% located in posterior fossa
  1) Optic chiasma / hypothalamus
  2) Cerebellum
  3) Brain stem

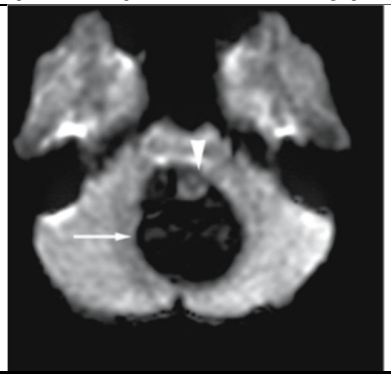
Imaging findings:

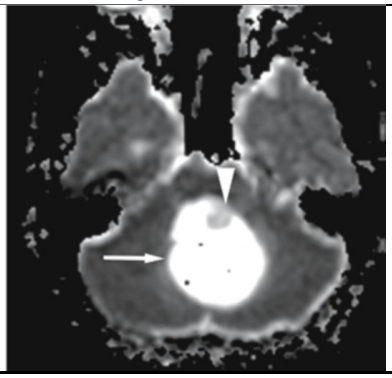
- Cystic lesion: CSF like signal
- Enhancing mural nodule / solid component





ADC map shows hyperintensity in the cystic component (arrow) and mild hyperintensity in the nodule > 1.2





5) Brain Stem Glioma

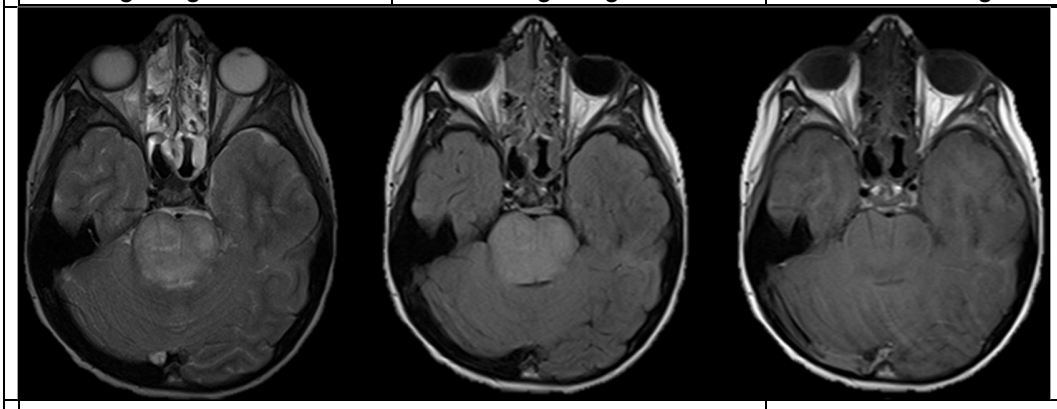
- 3rd common posterior fossa pediatric lesion
- Age : mean 10y
- Location:
  - Pons 50%
  - Medulla 30%
  - Midbrain
  - Tectal

❖ 2 types:

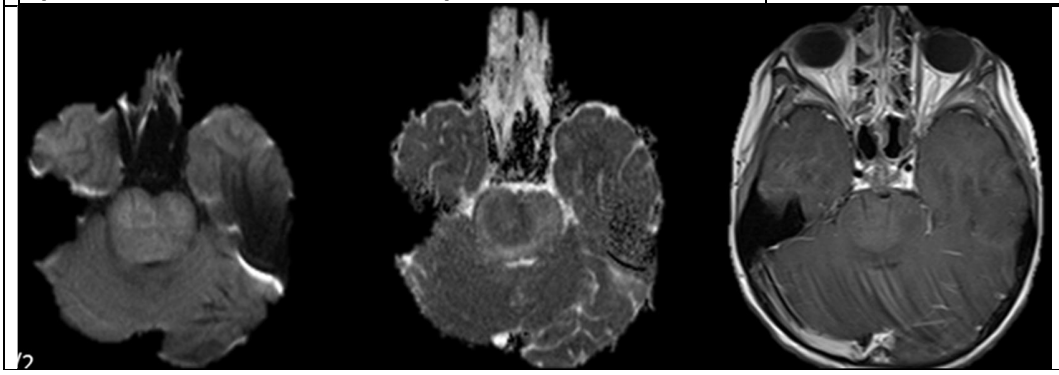
A) Fibrillary Astrocytoma (Grade II):

- Infiltrative non-enhancing lesion
- Diffusely expand brain stem
- Surround and engulf basilar artery
- Poor prognosis

T2: High signal	FLAIR: High signal	T1: iso-Low signal

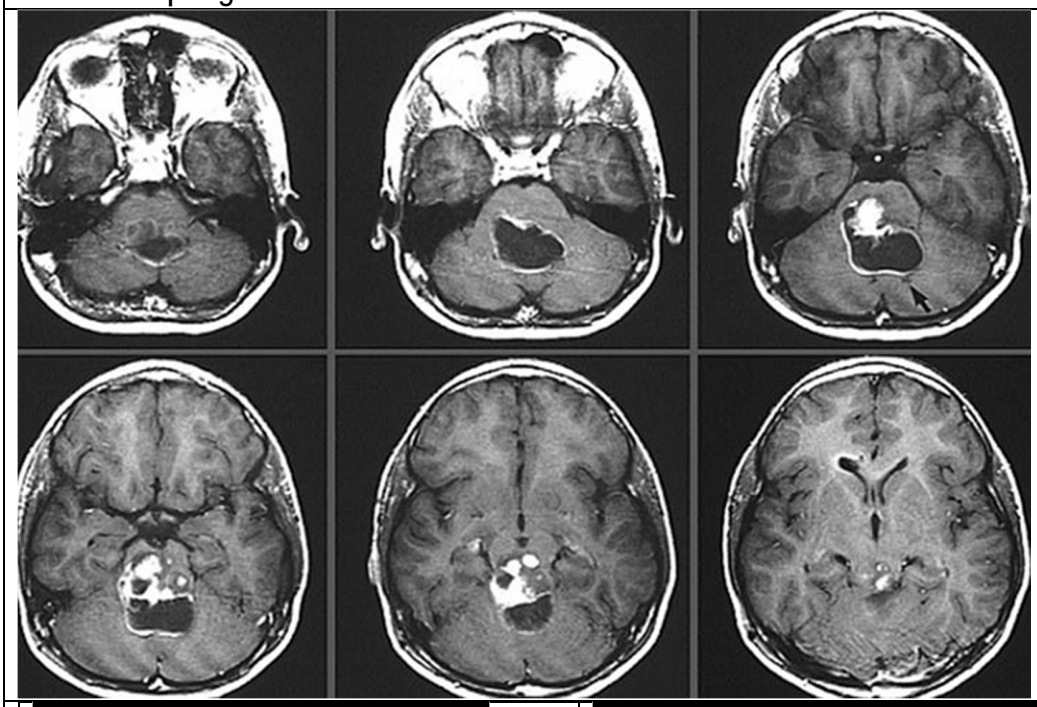


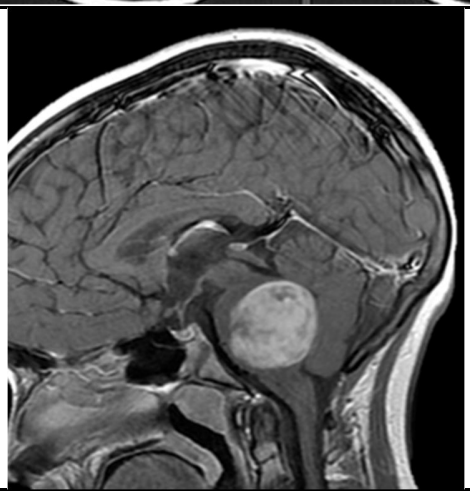
ADC: mild hyperintensity (0.85–1.17×10-3 mm2/s)	+C: No enhancement

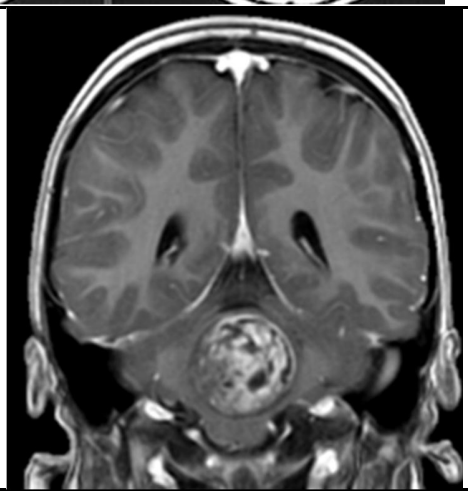


B) Pilocytic Astrocytoma (Grade I):

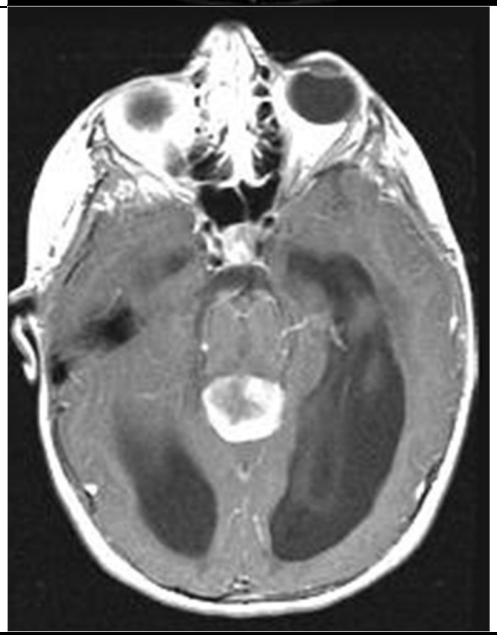
- Focal cystic lesion with enhancing mural nodule / solid component
- Exophytic component
- Anteriorly => encroaching on the prepontine / CPA cistern
- Posteriorly => encroaching on 4th ventricle => Obstructive hydrocephalus
- Better prognosis







❖ Tectal Glioma



V- Intraventricular Tumors

Septum Pellucidum

- Colloid Cyst
- Central neurocytoma
- SEGA (TS)
- Subependymoma

Trigone

Pediatric:

- Choroid plexus cyst
- Choroid plexus papilloma
- Choroid plexus carcinoma

Adult:

- Intraventricular Meningioma

4th Ventricle

Pediatric:

- Medulloblastoma
- Ependymoma
- Exophytic Brain stem glioma

Adult:

- Choroid plexus papilloma (solid enhancing)
- Subependymoma (Solid nonenhancing)
- Rosette-forming Glioneuronal Tumor (RGNT) (Cystic)

3rd Ventricle

- Septum pellucidum lesions

+

- Supra sellar/ hypothalamic lesions

+

- Pineal Lesions

+

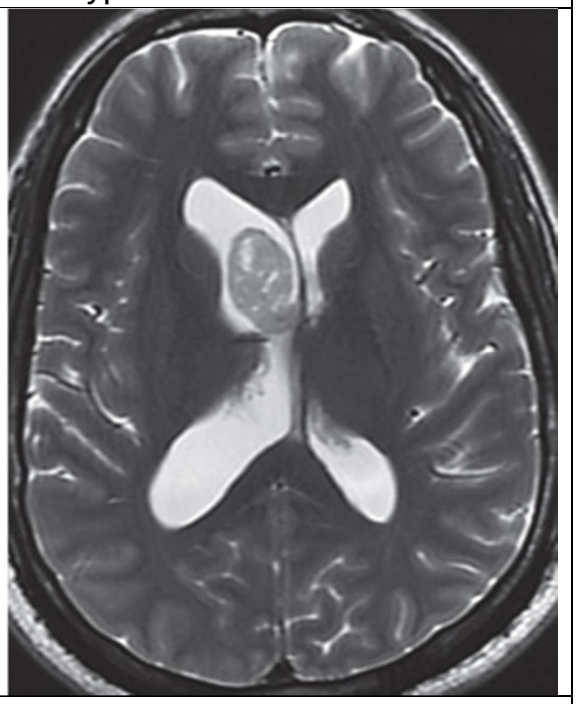
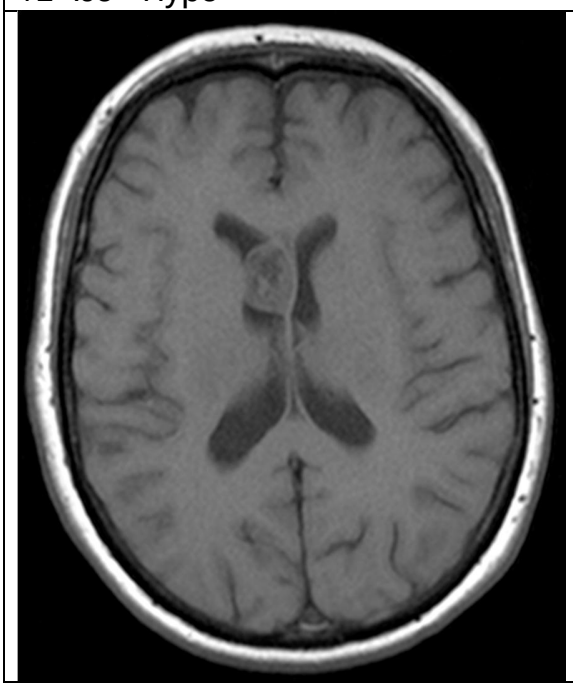
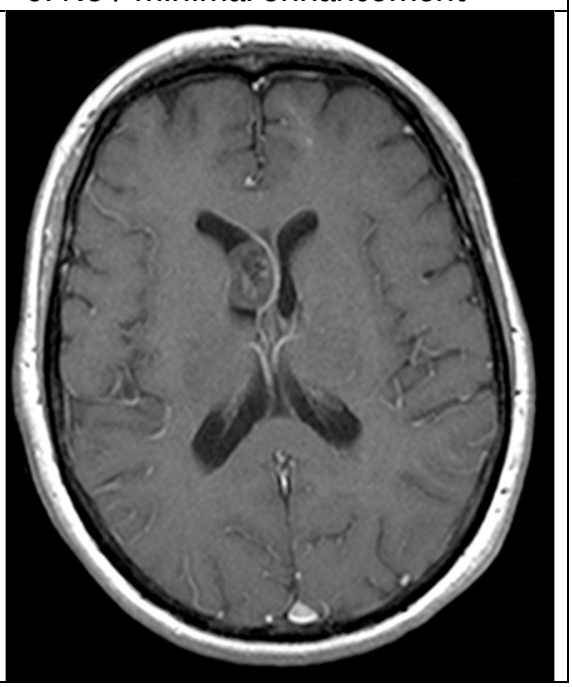
- Chordoid glioma

Diffuse Ependymal Lesions

- GM heterotopia
- TS subependymal noduels
- Ependymitis/ Ventriculitis
- CSF seedling:
  - –Intracranial tumor
  - –Extracranial tumor
- Lymphoma

A- Septum Pellucidum / Lateral Ventricle Lesions		
	1) Subependymoma	2) Central Neurocytoma
WHO	Grade I	Grade II
Incidence	0.2-0.7 %	0.25-0.5 %
	Arise from the subependymal glial layer surrounding the cerebral ventricles.	Arise from the septum pellucidum or ventricular wall.
Age	Middle age - Adult	
Location	60% 4th ventricle 40% Lateral ventricle	lateral ventricle with or without extension into the third ventricle
Imaging findings	•A well-circumscribed lesion. •Cystic degeneration. •Calcification rare. •Hhemorrhage rare.	• A well-circumscribed, lobulated masses • Multiple cyst like areas • "bubbly" appearance • 50% calcification • Hemorrhage => Intratumoral + Intraventricular
+C	•No/minimal enhancement. •No brain invasion. •No CSF dissemination	• Moderate / strong heterogeneous enhancement
	CT: hypo- to iso attenuating. T1: hypo- to isointense T2/ FLAIR: hyperintense	CT: hyperattenuating T1: isointense to gray matter T2: hyperintense

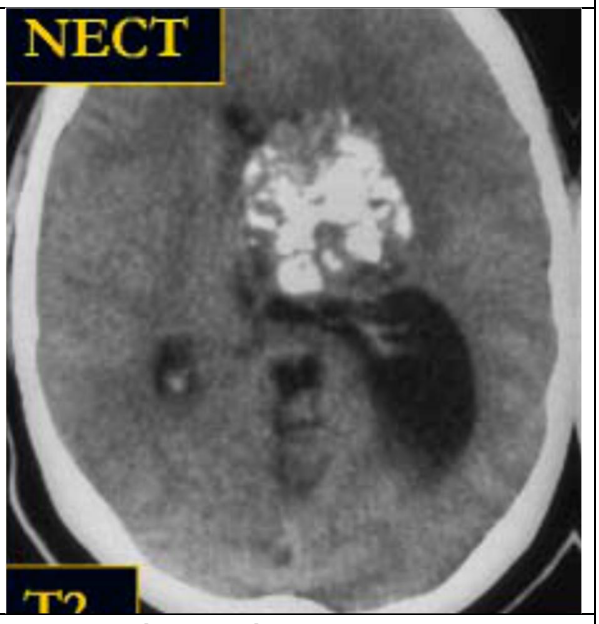
1) Septum pellucidum / Lateral ventricle
Subependymoma

CTL hypodense	T2: Hyperintense
	
T1: Iso - Hypo	+C: No / minimal enhancement
	

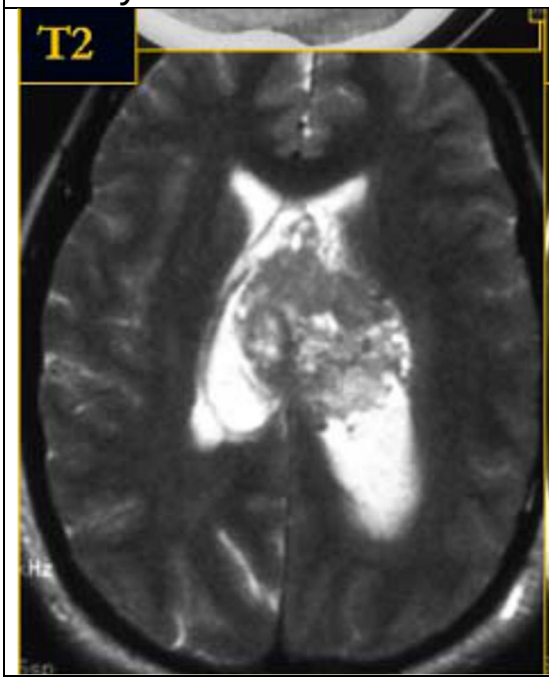
2) Central Neurocytoma

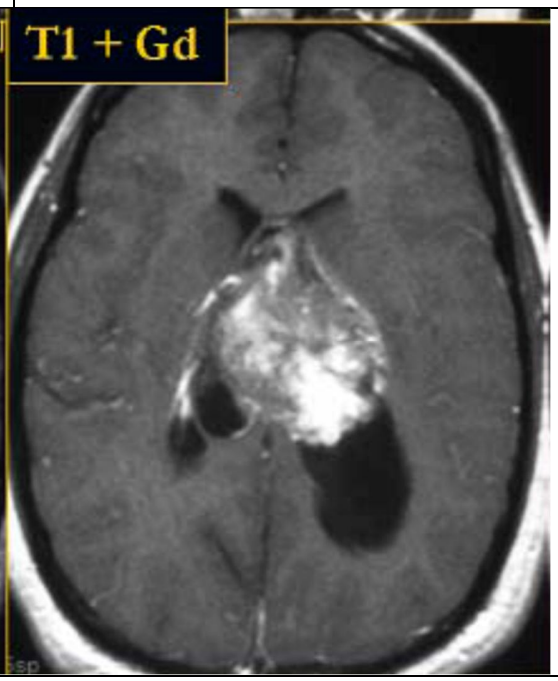
NCCT: Cystic components + Calcifications

T2: Heterogeneous high signal + Cystic components = Bubbly lesion



+C: Moderate heterogeneous enhancement



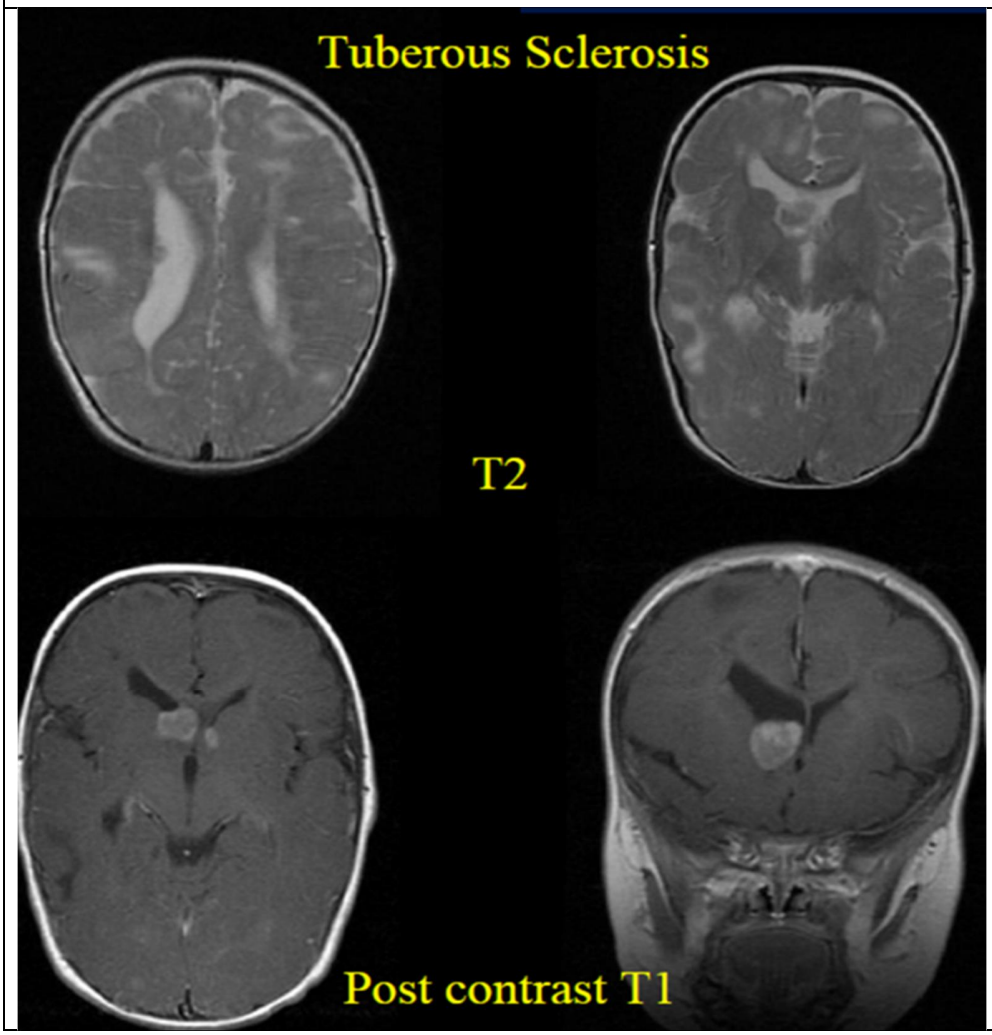


3) Subependymal Giant Cell Astrocytoma (SEGCA)

- *WHO grade I*
- is the most common cerebral neoplasm in patients with *tuberous sclerosis (TS)*, developing in up to 16% of cases.

- Location: arise near the foramen of Monro

- CT: hypo- to isoattenuating.
- T1: hypo- to isointense to gray matter
- T2: iso- to hyperintense
- T1+C: enhance

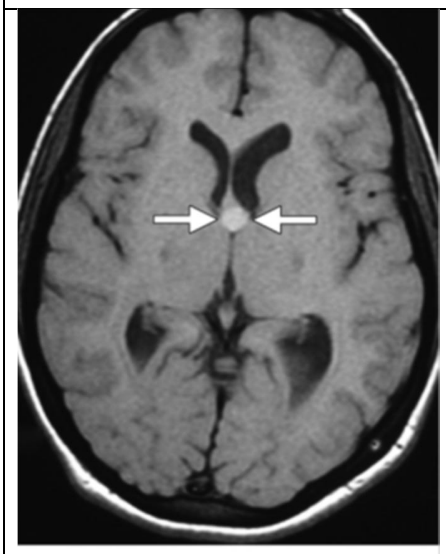


4) Colloid Cyst

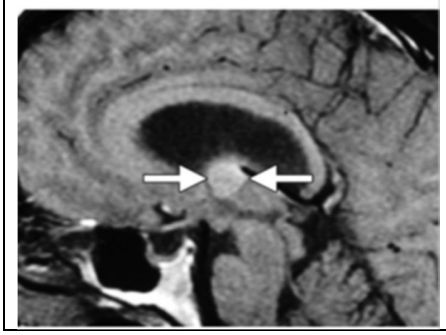
- A well defined small rounded lesion
- Centered on F. monro

Imaging findings:

- CT: hyperdense
- T1: high signal (high protencious content)
- T2: high signal
- +C: no enhancement



a.

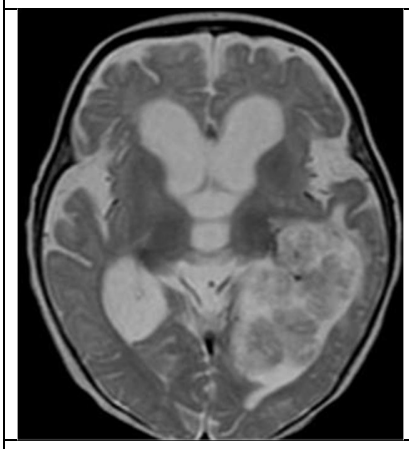


B- Atrium / Trigone Lesions		
	1) Choroid Plexus Papilloma	2) Intraventricular Meningioma
WHO	Choroid plexus papilloma (CPP) (WHO grade I) Atypical CPP (WHO grade II) Choroid plexus carcinoma (CPC) (WHO grade III)	
Incidence	5% of pediatric brain tumors	1-2% of intracranial meningioma
Age & location	*<5y: Trigone –3rd ventricle* *Adult: 4th ventricle - CPA*	*Adult: Atrium/Trigone of lateral ventricle*
	• Papillary or lobulated = Cauliflower appearance • Cystic areas • Calcifications • Flow voids	Well defined lesion Rare cystic changes Rare hemorrhage Common calcification
	CT: iso- to hyperattenuating T1: iso- to Low T2: iso- to High	CT: iso- hyperattenuating T1: iso- Low T2: iso- Low
+C	*Avid enhancement*	
	<u>Hydrocephalus:</u> CSF overproduction Mechanical obstruction	
	CPC => Drop metastasis	

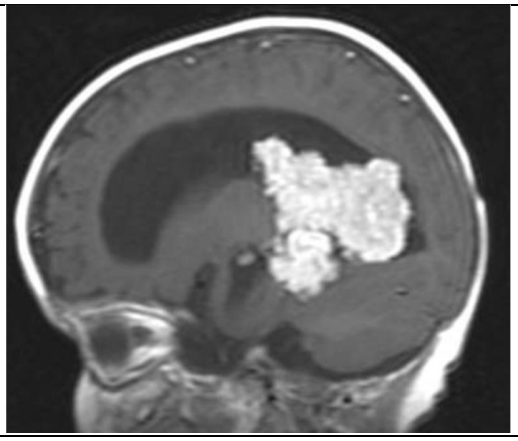
1) Choroid Plexus Papilloma

T2: Heterogeneous high signal

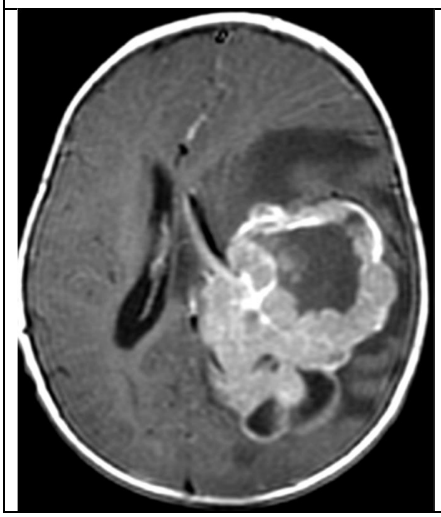
+C: Marked heterogeneous enhancement

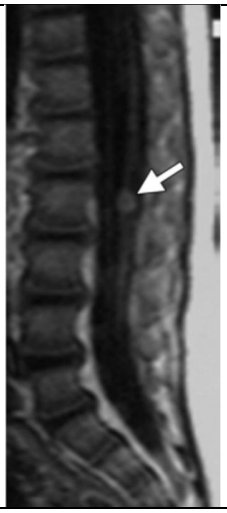


Choroid Plexus Carcinoma

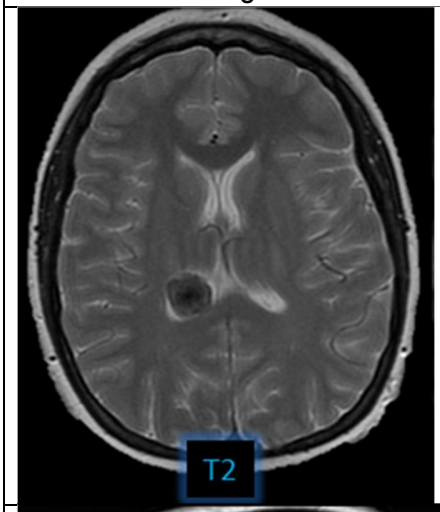
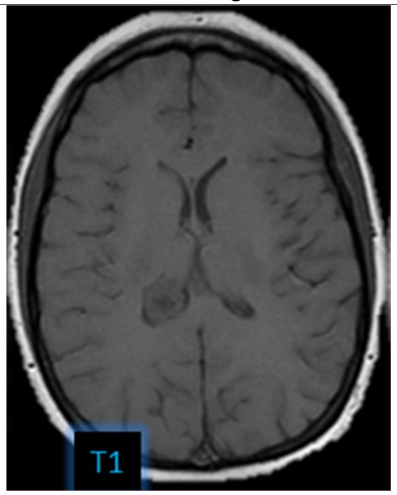
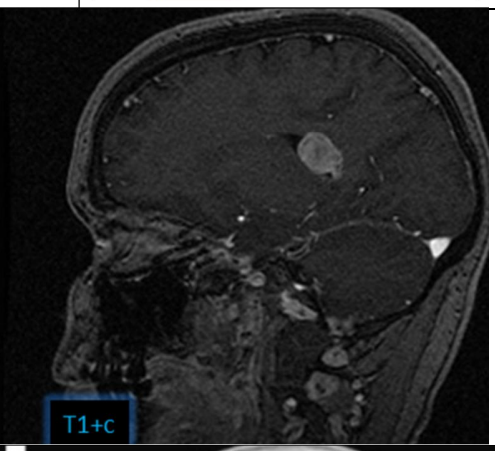
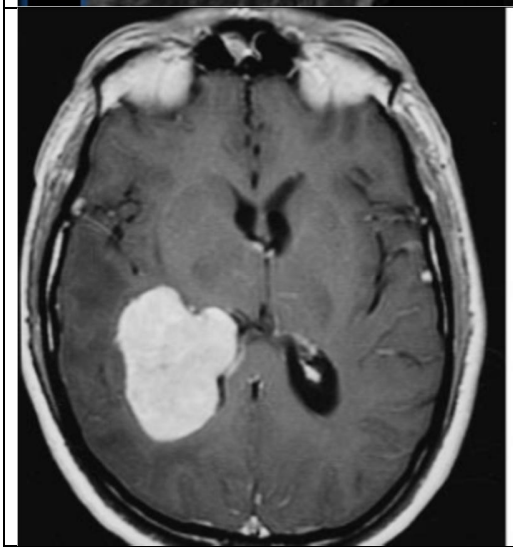
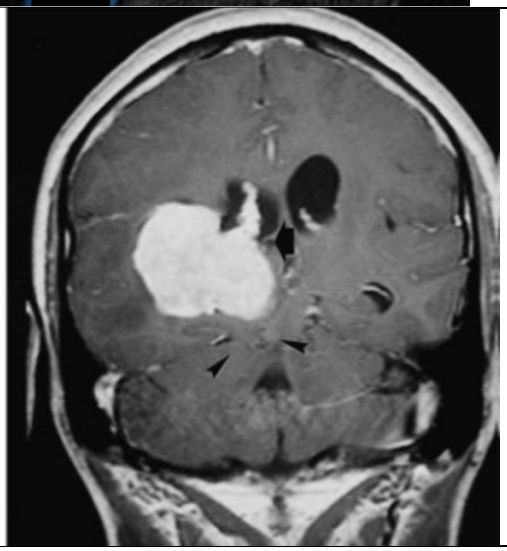


CSF Dissemination



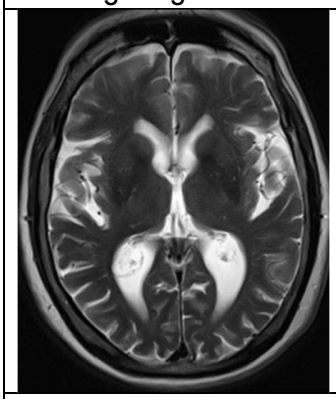
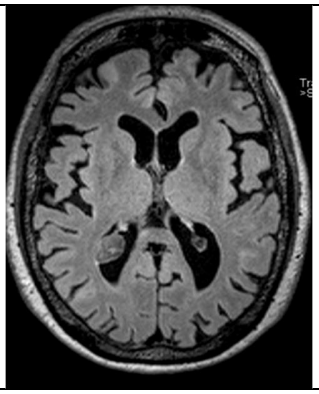
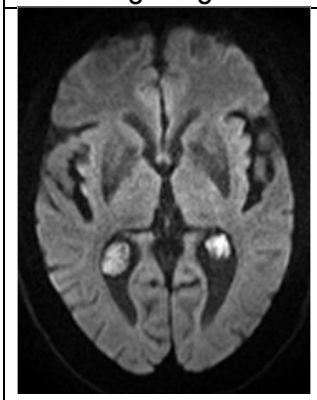
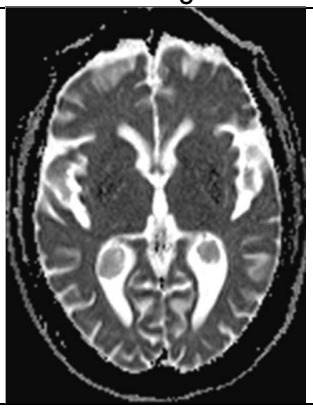
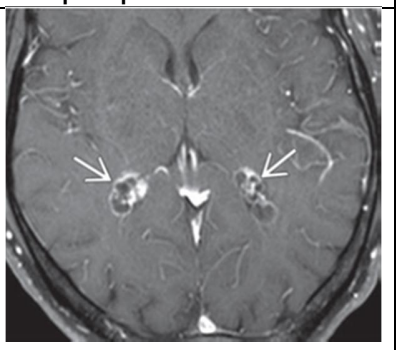


2) Intraventricular Meningioma

T2: iso – Low signal	T1: Iso – Low signal
	
	
	

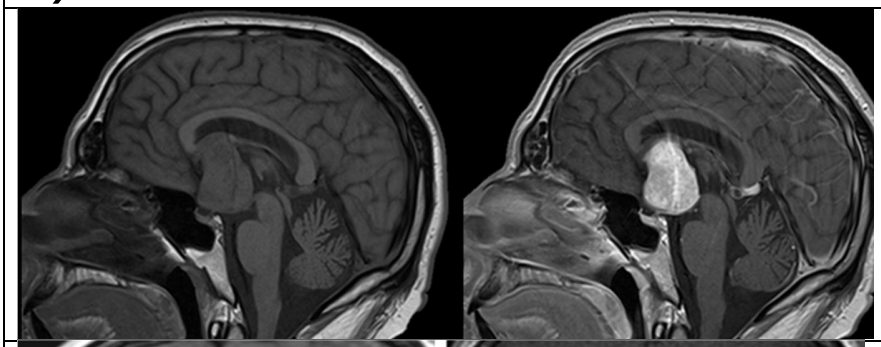
	3) Choroid Plexus Xanthogranuloma	4) Choroid Plexus Cyst
Incidence	Common, incidental and almost asymptomatic lesions	
	Bilateral in 2/3 of cases	
Location	*Trigone of lateral ventricle*	
	mixture of lipid, fluid and blood products	Pure fluid CSF like signal
CT	*Peripheral choroid plexus Calcification*	
Imaging findings	T1: Variable SI T2: High SI FLAIR: not fully suppressed DWI: High signal	T1: Low T2: High FLAIR: Low DWI: Low
+C	*No / Rim / Nodular enhancement*	

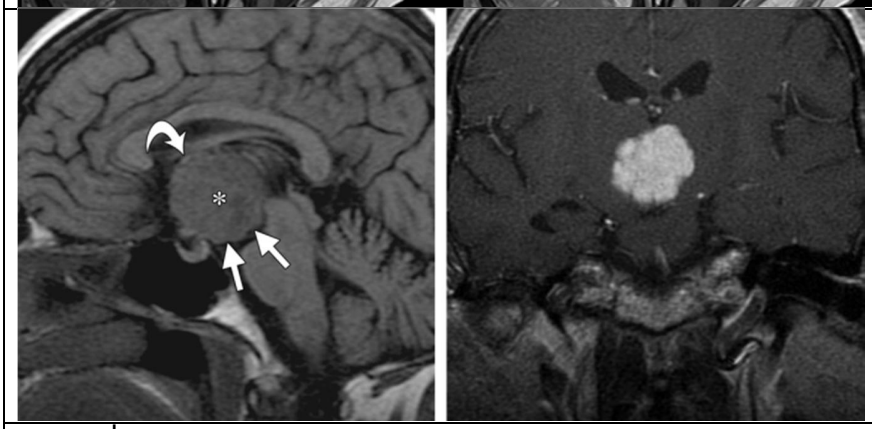
Choroid Plexus Xanthogranuloma

T2: High signal	FLAIR: not fully suppressed	
		
DWI: high Signal	ADC: Low signal	+C: peripheral nodular
		

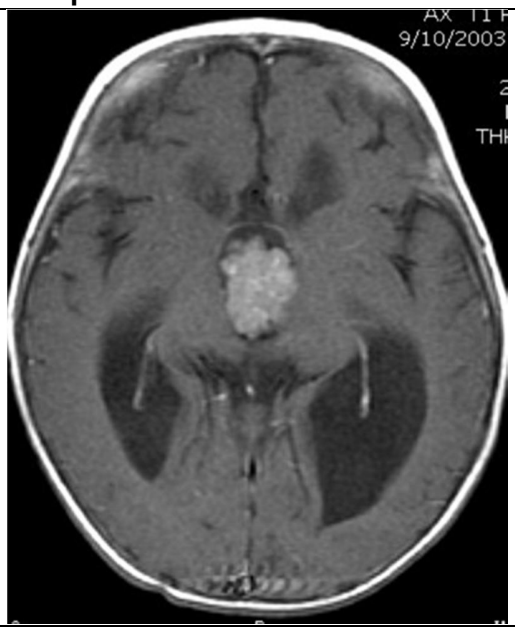
C- 3rd Ventricular Lesions		
	1) Chordoid Glioma	2) Choroid Plexus Papilloma
WHO	Grade II	Choroid plexus papilloma (CPP) (WHO grade I) Atypical CPP (WHO grade II) Choroid plexus carcinoma (CPC) (WHO grade III)
Incidence	Rare	5% of pediatric brain tumors
Age		<5y: Trigone –3rd ventricle
	A well-defined ovoid mass Arises from anterior 3rd ventricle Adherent to hypothalamus	•Papillary or lobulated = Cauliflower appearance •Cystic areas •Calcifications •Flow voids
	CT: iso- to hyperdense T1: iso - Low signal T2: Iso – High signal	CT: iso- to hyperdense T1: iso- to Low T2: iso- to Low
+C	T1+C: homogenous intense enhancement	

1) Chordoid Glioma





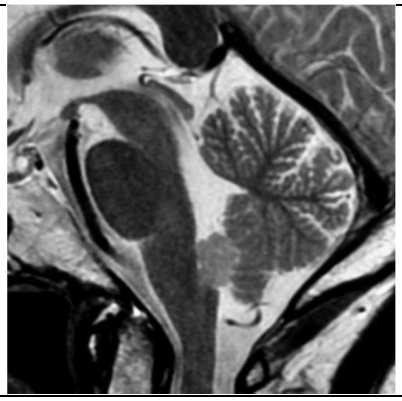
2) 3rd Ventricle Choroid Plexus Papilloma



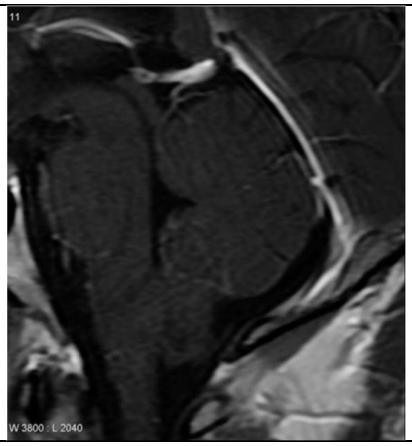
D- Adult 4th Ventricular Lesions		
	Subependymoma	Choroid Plexus Papilloma
WHO	Grade I	Choroid plexus papilloma (CPP) (WHO grade I) Atypical CPP (WHO grade II) Choroid plexus carcinoma (CPC) (WHO grade III)
Incidence	0.2%–0.7% of brain tumors	5% of pediatric brain tumors
Age	Middle age - Adult	<5y: Trigone –3rd ventricle Adult: 4th ventricle - CPA
Location	60% 4th ventricle 40% Lateral ventricle	
	•A well-circumscribed lesion. •Cystic degeneration. •Calcification rare. •Hemorrhage rare.	•Papillary or lobulated = Cauliflower appearance •Cystic areas •Calcifications •Flow voids
	CT: Hypo- to iso. T1: Hypo- to iso T2: High	CT: Hypo- to iso. T1: Hypo- to iso T2: High
	No or minimal enhancement. No invasion into brain. No CSF dissemination	*Marked heterogeneous enhancement*

1) 4th Ventricle Subependymoma

T2/ FLAIR: High signal



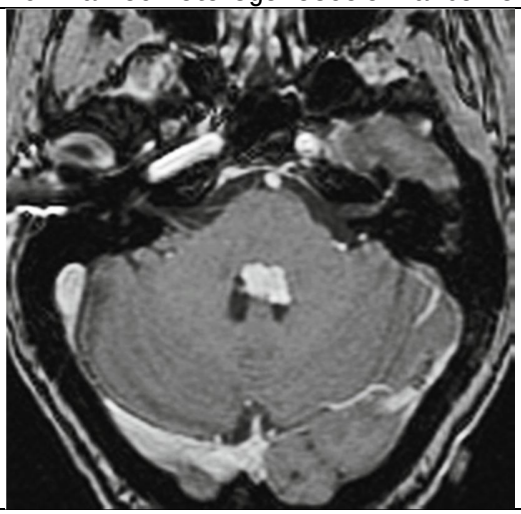
+C: No enhacement



2) 4th Ventricle Choroid Plexus Papilloma

T2/FLAIR: high signal

+C: Marked heterogeneous enhancement

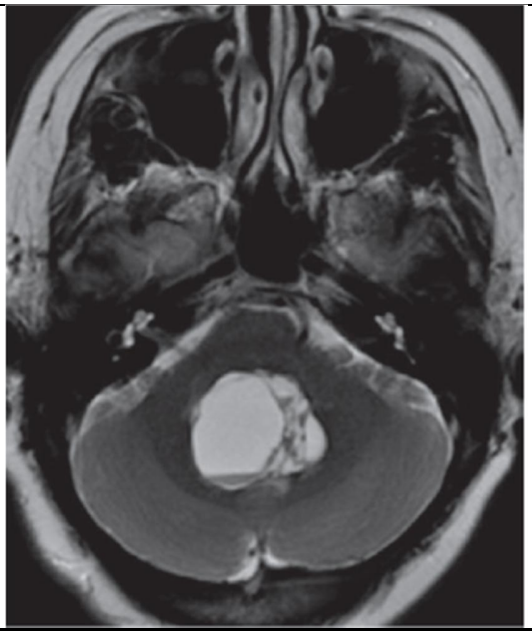


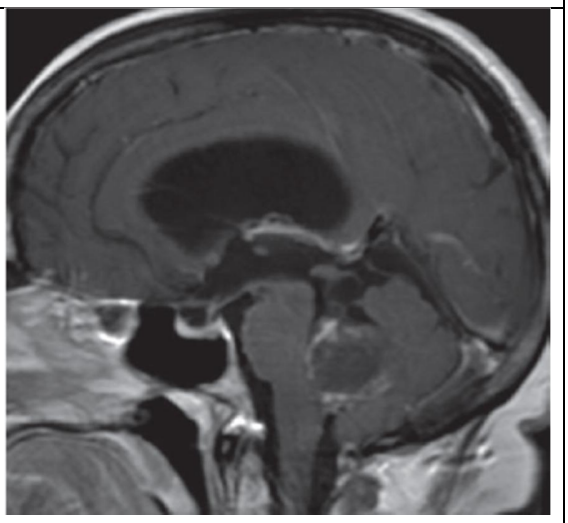
3) Rosette-forming Glioneuronal Tumor

- WHO grade I
- Rare primary brain tumor.
- Age: Young Adults
- Location: 4th ventricle

Imaging findings:

- Cystic neoplasm centered on 4th ventricle
- Intratumoral hemorrhage and calcification may occur





E- Intra-ventricular Cystic Lesions

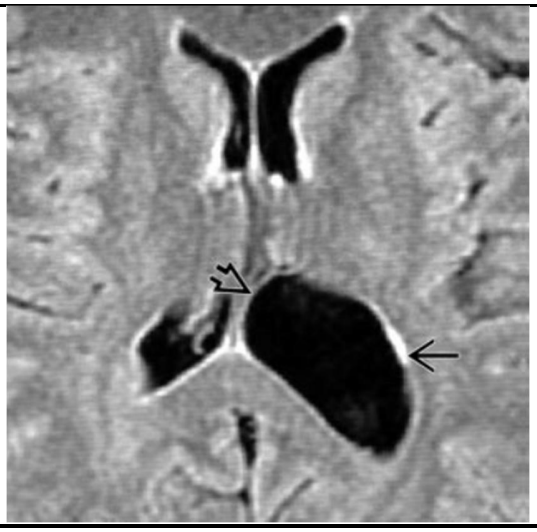
- Cavum septum pellucidium *(CSP)*
- Cavum vergae *(CV)*
- Cavum veli interpositi *(CVI)*
- Choroid plexus cyst
- Choroid plexus xanthogranuloma
- Arachnoid cyst
- Ependymal cyst
- Colloid cyst
- Trapped ventricle by tumor

Ependymal Cysts

- Rare, benign, ependymal-lined cysts
- Location: Lateral ventricle or juxtaventricular region of the temporoparietal region and frontal lobe

Imaging findings:

- CSF like signal
- Lateral displacement of the choroid plexus



VI- Extra-Axial Lesions

Specific Locations
A. Sellar / Suprasellar / Hypothalamic
B. Parasellar / Cavernous Sinus
C. Clival / Basisphenoid / Foramen magnum
D. Pineal / Quadrigeminal cistern
E. Petrous & CPA/ IAC & Jugular foramen (See petrous)
General Lesions
F. Subarachnoid Lesions
G. Meningeal Lesions
H. Bone Lesions

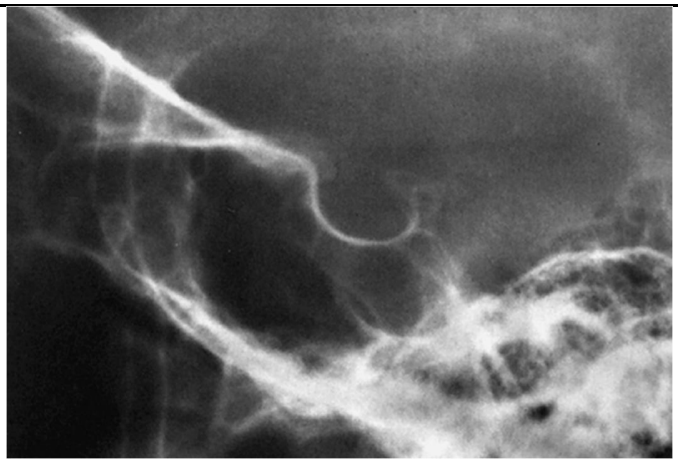
A- Sellar / Suprasellar / Hypothalamic / Pituitary Lesions

❖ Sella

Concave midline depression in basisphenoid

<u>Anterior border:</u>
Tuberculum sellae
Anterior clinoid processes

<u>Posterior border:</u>
Dorsum sellae
Posterior clinoid processes



❖ Pituitary Gland

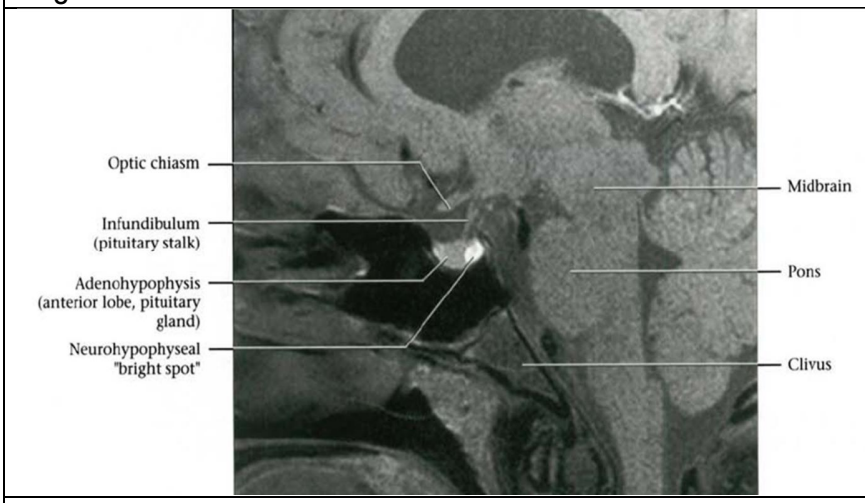
<u>Adenohypophysis = Anterior Lobe (80%)</u>
PRL (10%-30%)
GH (50%)
ACTH (10%-30%)
TSH (5%)
FSH/LH (10%)

<u>Neurohypophysis = Posterior Lobe (20%)</u>
Vasopressin (ADH) & Oxytocin
High T1 signal = bright spot
Not suppressed by Fat suppression sequences

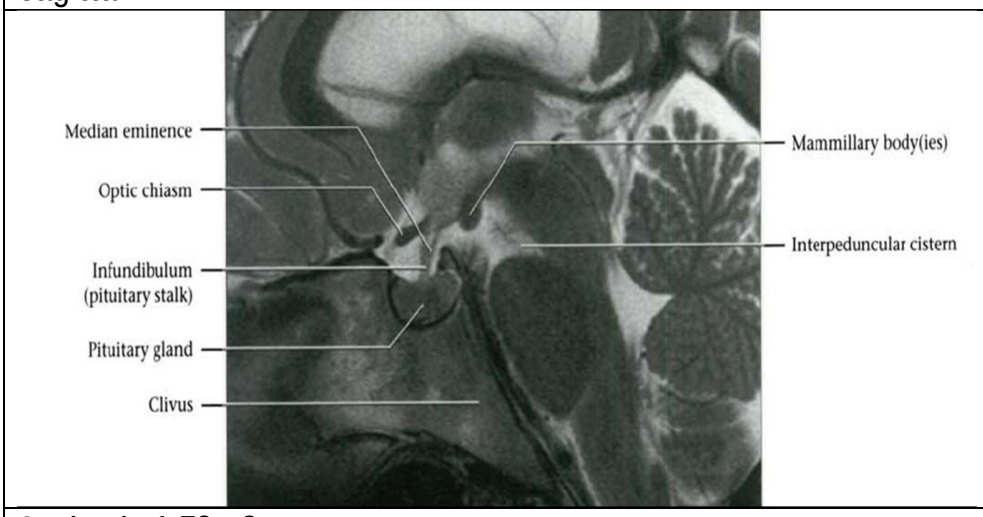
<u>Height (in the midsagittal plane):</u>
- ≤ 6 mm in children
- 8 mm in males and post menopausal women.
- 10 mm physiologic hypertrophy in young females
- 12 mm in pregnant/lactating females

<u>Superior border</u>
- Straight or slightly concave
- Convex only during puberty or pregnancy

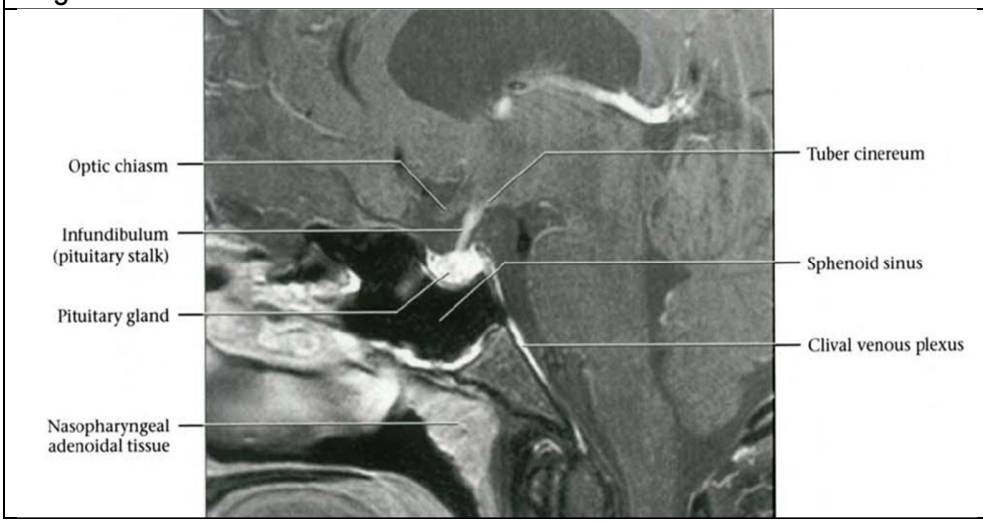
Sagittal T1 FS



Sagittal T2

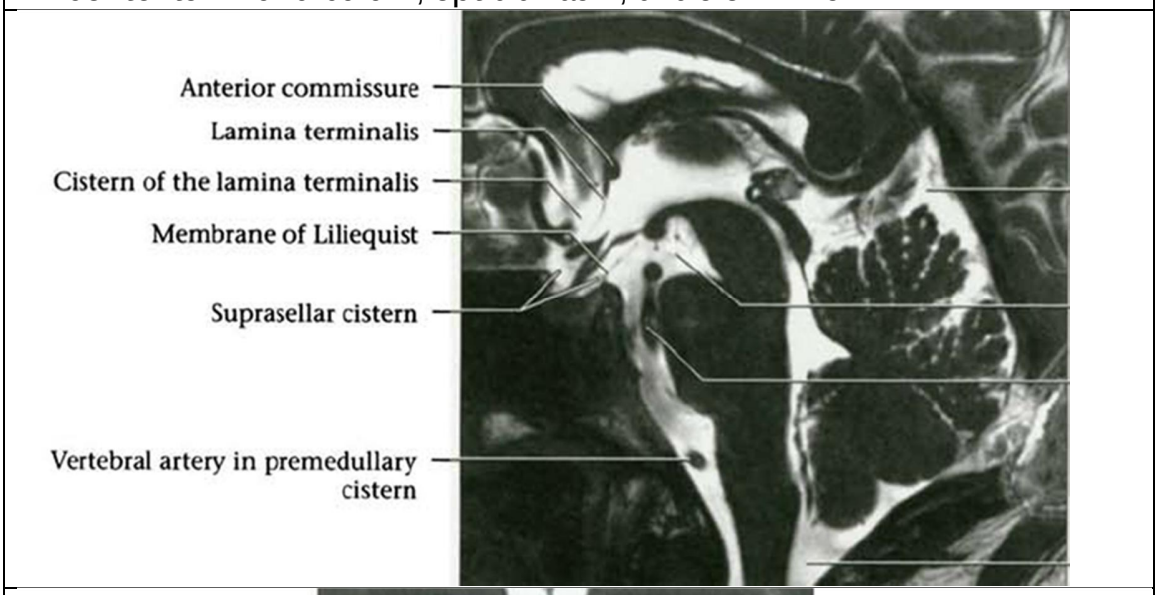


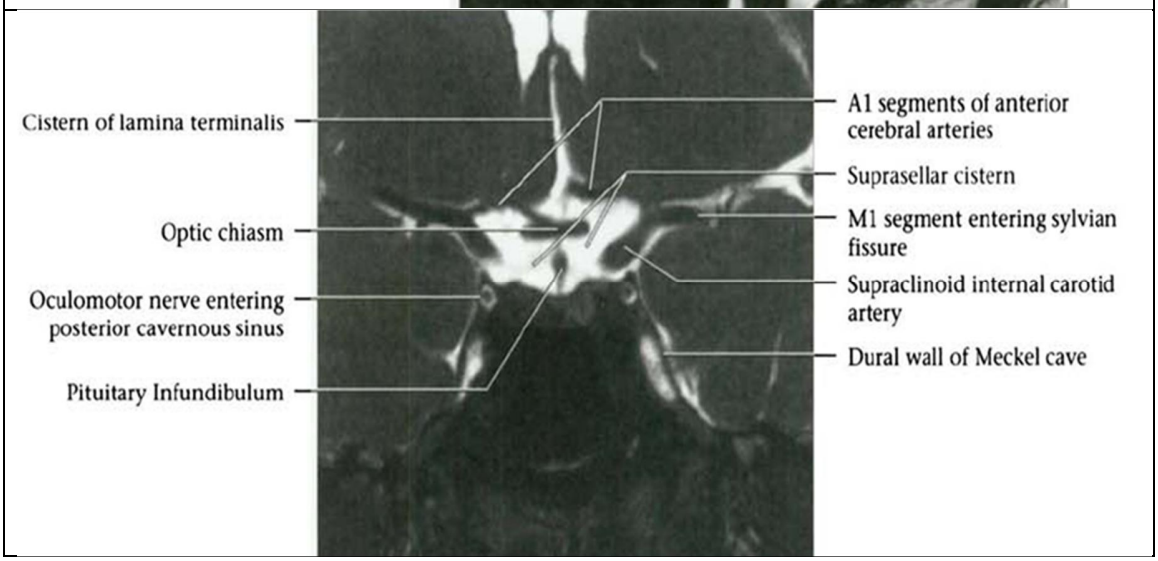
Sagittal T1 FS +C



❖ Supra-Sellar Cistern

- Superior to pituitary gland
- Contents: Infundibulum, optic chiasm, circle of Willis




Cistern of lamina terminalis
Optic chiasm
Oculomotor nerve entering posterior cavernous sinus
Pituitary Infundibulum
A1 segments of anterior cerebral arteries
Suprasellar cistern
M1 segment entering sylvian fissure
Supraclinoid internal carotid artery
Dural wall of Meckel cave

❖ Infundibulum

- Thin stalk tapers smoothly from the floor of the third ventricle to the pituitary gland

<u>Normal infundibulum width:</u>

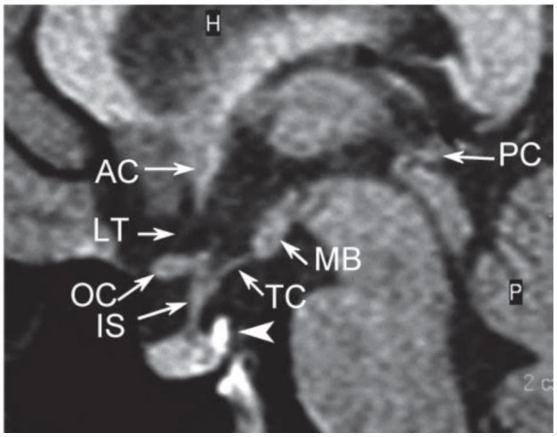
- 3 mm at its origin
- 2 mm near its insertion

- Midline

- It enhances after the administration of intravenous contrast medium.

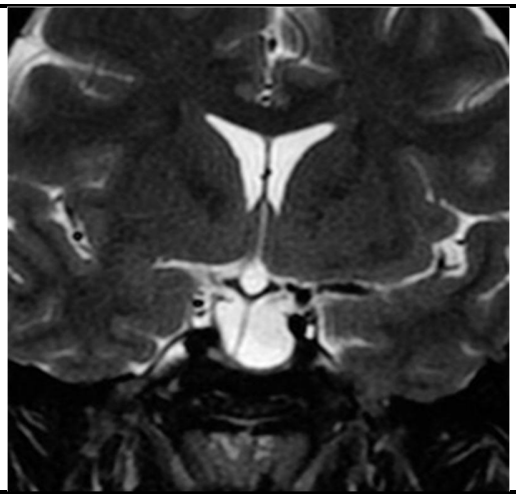
❖ Hypothalamus

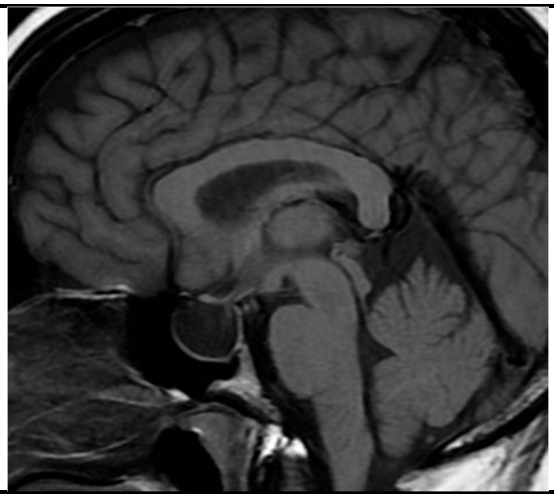
- Anterior commissure (AC)
- Posterior commissure (PC)
- The anterior boundary of the hypothalamus is the lamina terminalis (LT), which extends between the optic chiasm (OC) and the anterior commissure
- The posterior boundary is imprecise; it is indicated by a line that extends between the mamillary bodies (MB) and the posterior commissure.
- The floor of the hypothalamus is formed by the infundibular stalk (IS), the tuber cinereum (TC), and the mamillary bodies.



❖ Empty Sella

- Normal variant when seen in isolation
- Rarely symptomatic (may be associated with pseudotumor cerebri = idiopathic intracranial hypertension)
- Protrusion of arachnoid, CSF into sella
- Normal pituitary becomes flatted, displaced posteroinferiorly against sellar floor



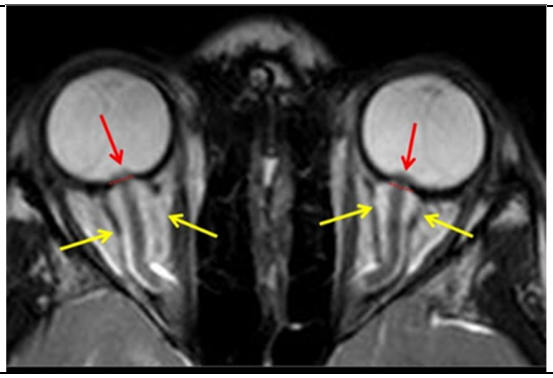


❖Idiopathic Intracranial Hypertension = Pseudotumor Cerebri

- A syndrome associated with elevated CSF pressure, visual changes, and headaches
- Typically seen in obese black females.

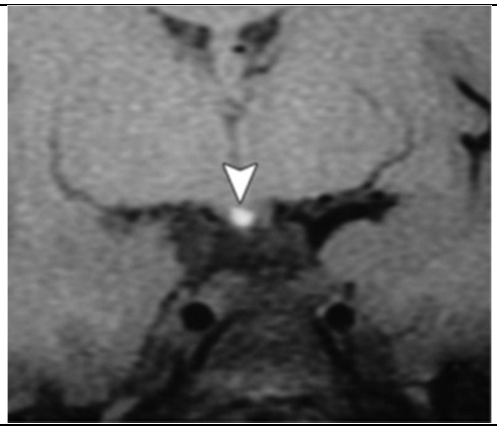
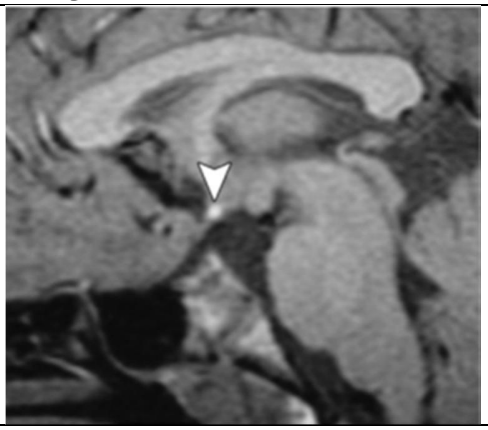
<u>Imaging findings include:</u>

- Empty sella
- Enlargement of Meckel's cave,
- Optic disc protrusion into the posterior globes.
- The ventricles are slightly reduced in caliber.
- The sigmoid or transverse sinus may be stenotic.

<u>Papilledema:</u> Flattening of posterior sclera Protrusions of optic nerve heads Vertical tortuosity of optic nerves Prominent subarachnoid space around the optic nerves	

❖ Ectopic Posterior Pituitary

- Posterior lobe: high T1 SI
- May be ectopic, beneath infundibulum
- Congenital or acquired, especially post-traumatic

Coronal T1	Sagittal T1
	

❖Origin Of Sellar And Suprasellar Region Lesions

- Pituitary gland:
  - Microadenoma
  - Macroadenoma
  - Pituitary Apoplexy
  - Rathke-Cleft cyst
  - Craniopharyngioma
- Optic chiasma
  - Chiasmatic glioma
- Hypothalamus
  - Glioma
  - Hamartoma
  - Germinoma
- Infundibulum
  - Granulomatous hypophysitis
    - TB / Sarcoidosis / Wegner granulomatosis / LCH
  - Meningeal carcinomatosis
  - Lymphocitic hypophsitis
- Meninges
  - Planum sphenoidale Meningioma
  - Granulomatous meningitis
  - Meningeal carcinomatosis
- Sphenoid sinus
  - Sinusitis / fungal
  - Mucocele
  - Sphenoid sinus tumors
- Skull base = Basisphenoid / Clival
  - 1ry : Chordoma - Chondrosarcoma
  - 2ry : Metastasis
- Nasopharyngeal
  - Nasopharyngeal carcinoma
  - Nasopharyngeal angiofibroma
  - Nasopharyngeal RMS

i- Pediatric Sellar and Suprasellar Lesions

	Craniopharyngioma (adenanatous type)	Glioma (Chiasmatic – hypothalamic)	Hypothalamic Hamartoma	Germinoma
Age	2 peaks Child < 20 Y Middle age	Child < 20, Associated with *NF-1*	Childhood M > F	Childhood 10-20 Y
Location	Predominant supra-sellar with smaller intrasellar component	*Supra sellar*	Suprasellar Sessile or peduculated mass in *Tuber cinerum* (between infundibulum & mamillary bodies)	*20% supra sellar* (infundibulum hypo thalamus) *80% pineal body*
CT	Lobulated 90% cystic / solid 90% calcification .	iso dense no cystic component no calcification no hge	rounded iso dense no cystic component No calcification No hge	midline mild hyper dense no cystic component No hge
MRI	Solid :- hypo in T1 hyper in T2 Cystic:- hyper T1 hyper T2	T1 : Iso T2 : Hyper	T1 : Iso T2 : Hyper	T1 : Iso T2 : Iso
+C	Strong enhancement of solid portion & periphery	Strong homogeneous enhancement	*NO enhancement*	Strong homogeneous enhancement
Spread	Displaces pit. Stalk posteriorly	Infiltrate thalamus , Brain stem & Visual tracts	Extend to interpeduncular c. or bulge into 3rd ventricle	

1) Craniopharyngioma

- Arise from remnant of craniopharyngeal duct

2 pathology:

- Adamantinomatous (children) type
- Papillary (adults) type

2 peaks:

- Children < 20 y
- Adults: 4th - 6th decade of life

Location:

- 75% Suprasellar
- 21% Sellar and suprasellar
- 4% Intra-sellar

Imaging Findings:

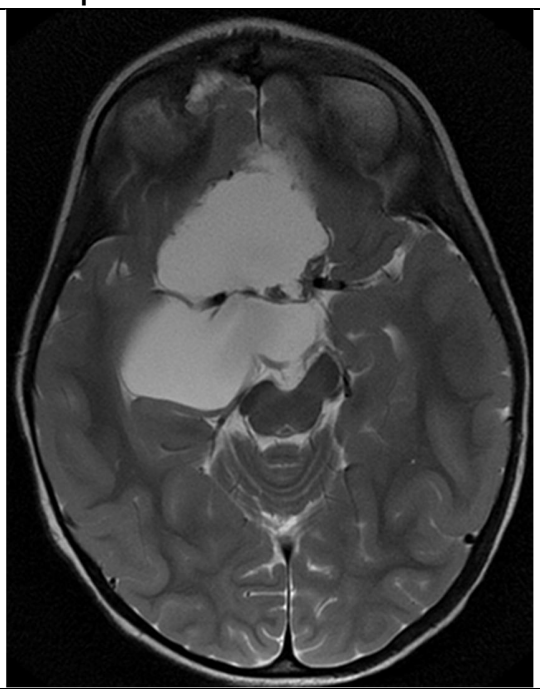
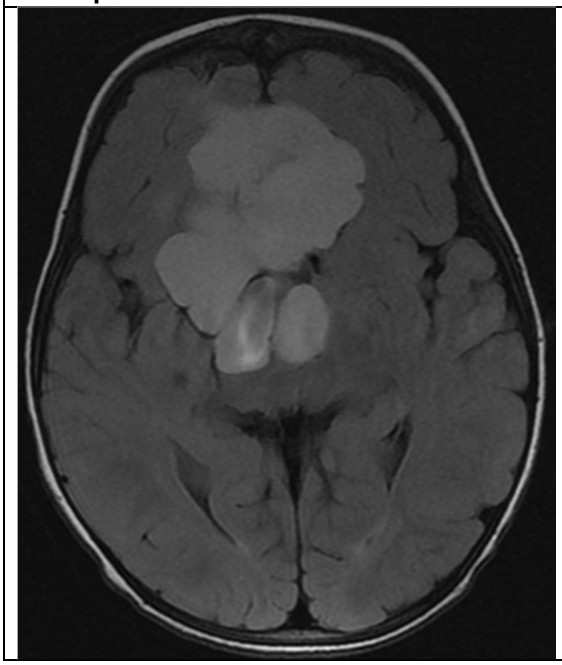
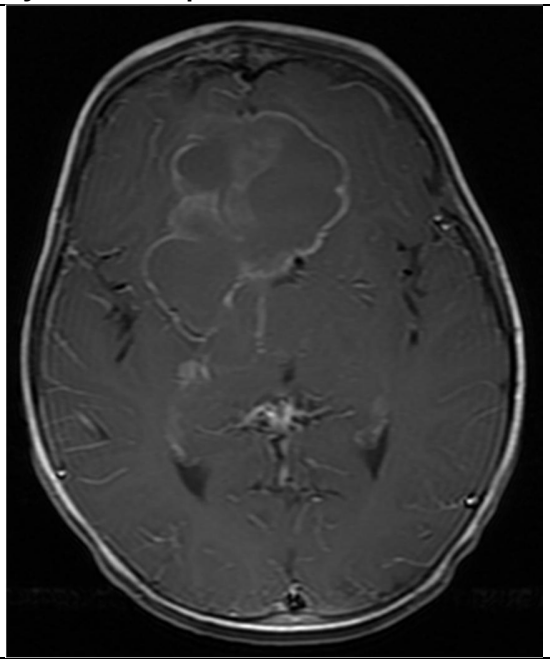
- 90% Cystic and solid components.
- 90% Calcification
- Occupies anterior of 3rd ventricle
- 2ry dilatation of the lateral ventricles

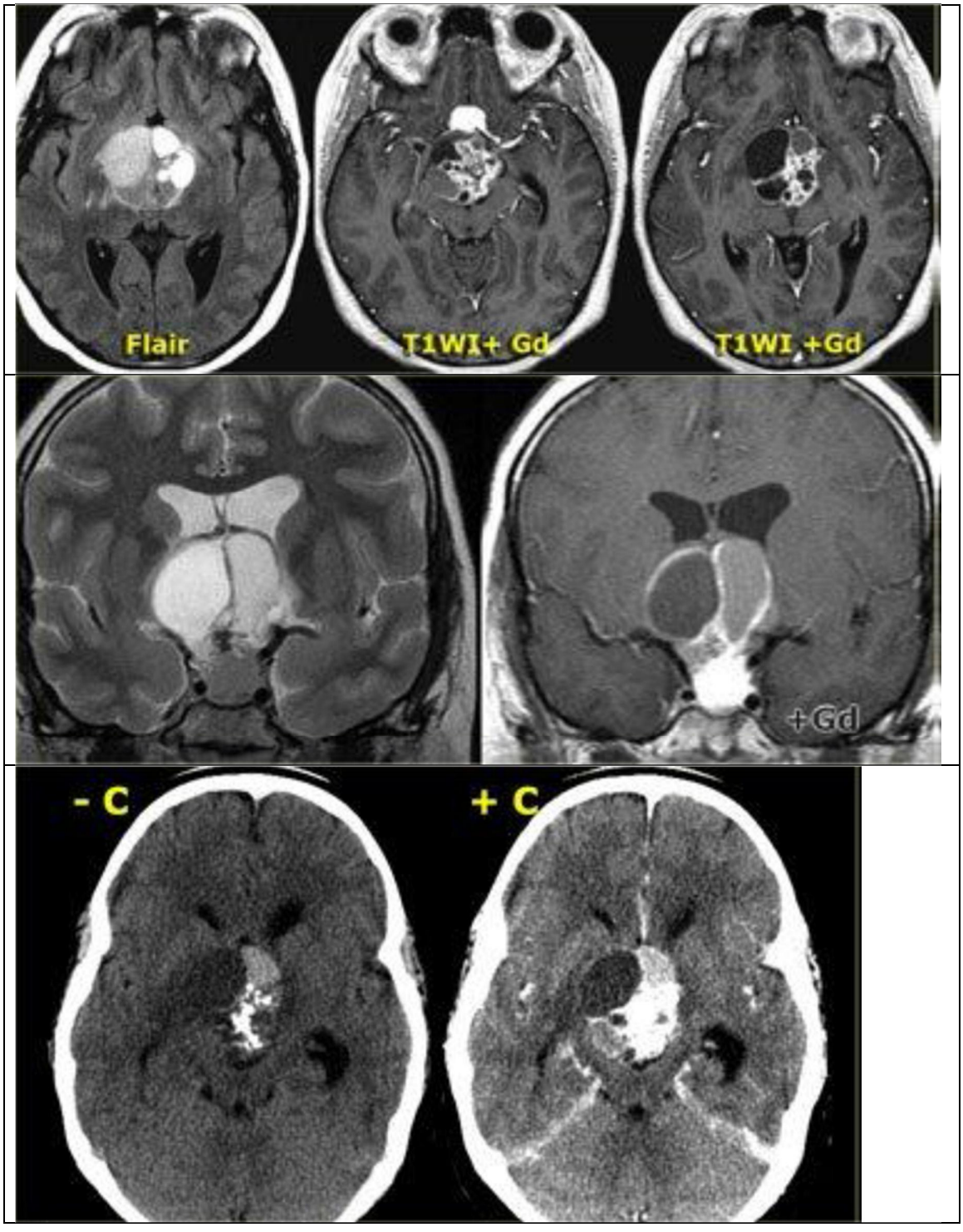
The cystic component:

- T1: Low or high signal
- T2: High signal
- +C: Rim enhancement

The solid component:

- T1: Low signal
- T2: High signal
- +C: Heterogeneous enhancement

CT: hypodense cystic component + Calcification	T2: High signal of cystic component
	
FLAIR: High signal of cystic component	+C: Marginal enhancement of cystic component
	


Flair
T1WI+ Gd
T1WI +Gd
+Gd
- C
+ C

2) Optic Pathway/Hypothalamus Glioma

- Arises from optic pathway (optic nerve / chiasma/ tracts) or hypothalamus.
- 30-50% have family history of NF1

- Age = 2-4yrs age
- Sex: M=F

Clinical picture:

- Hydrocephalus, decreased vision, Pituitary dysfunction (short stature)

2 Pathology:

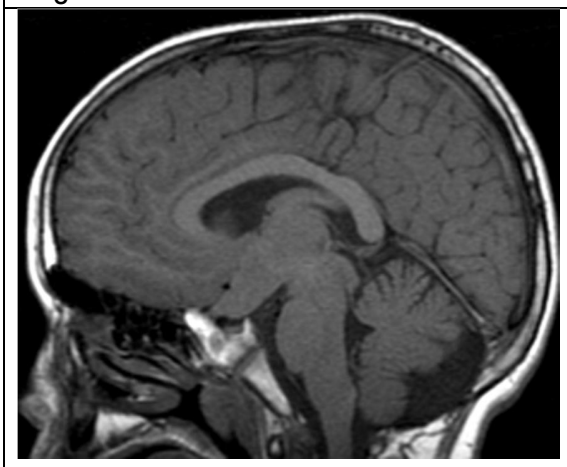
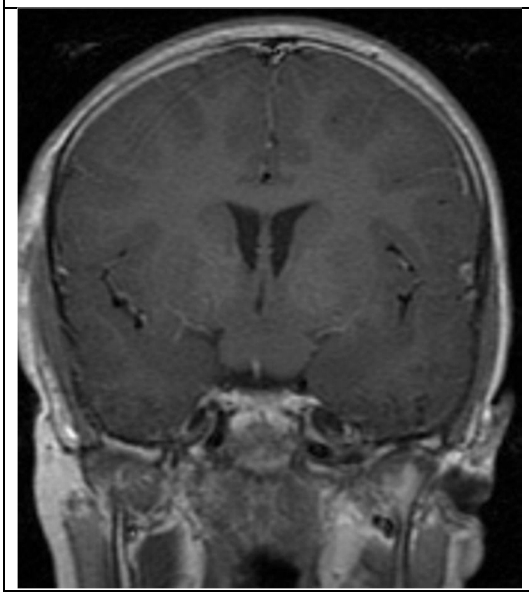
- 60% JPA (WHO Grade I)
- 40% Fibrillary (WHO grade II)

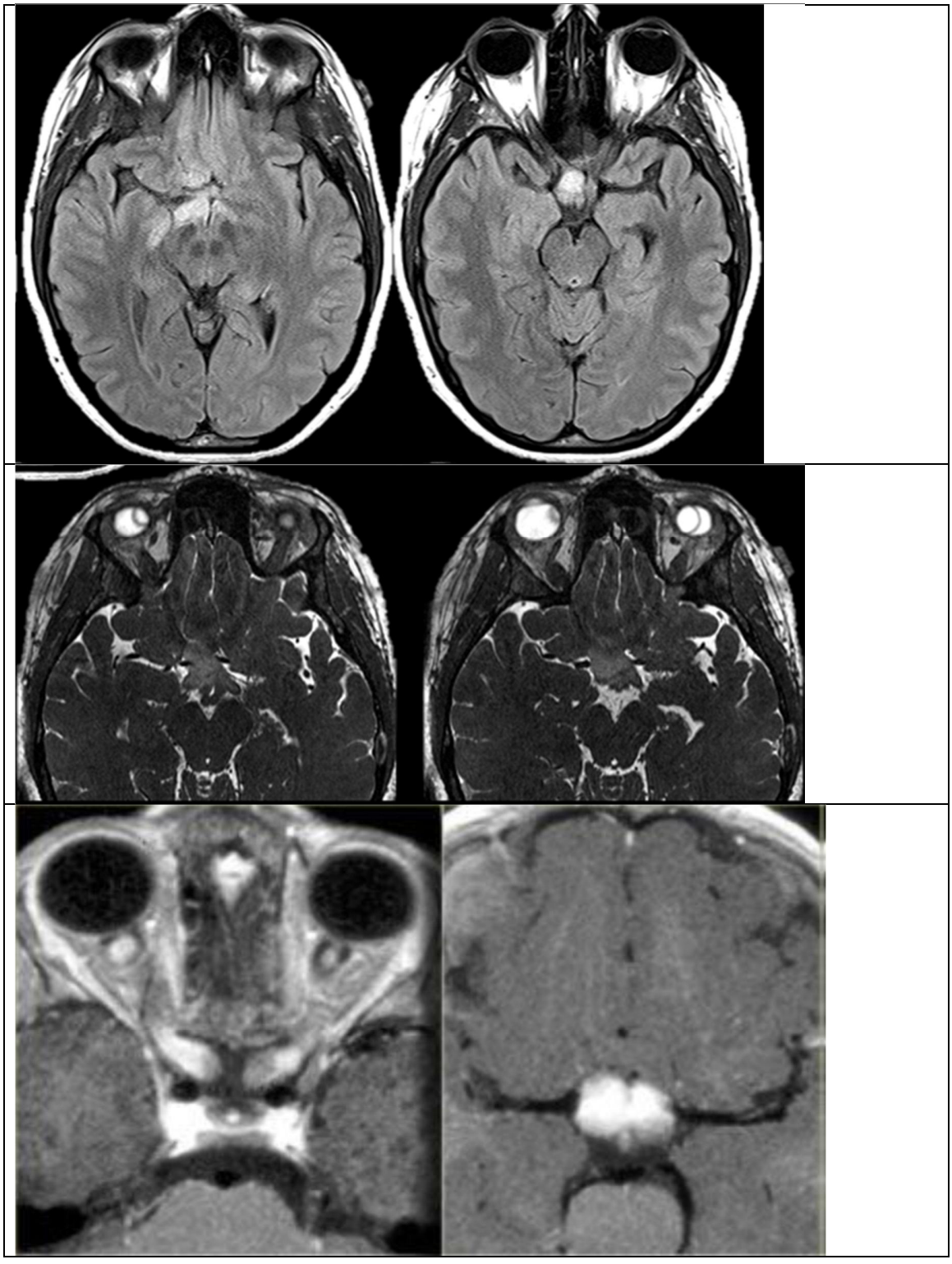
3 Subtypes:

- Child with NF-1:
  - Bilateral anterior optic pathways
  - Extends to optic chiasma and retrochiasmatic
- Child without NF-1:
  - Predominantly optic chiasma and retrochiasmatic
- Adult:
  - Unilateral optic nerve with posterior extension

Imaging findings:

- Suprasellar lesion
- Occupies anterior of 3rd ventricle.
- 2ry dilatation of lateral ventricles.
- Fusiform or lobulated enlargement of the optic pathway
- No / mild enhacement

Sagittal T1	Axial T2
	
Coronal T1 +C	**Axial T1+C**
	



3) Suprasellar Germinoma

- Originate in hypothalamus and extend into infundibulum

- Age: children and Adolescents
- Sex: M=F

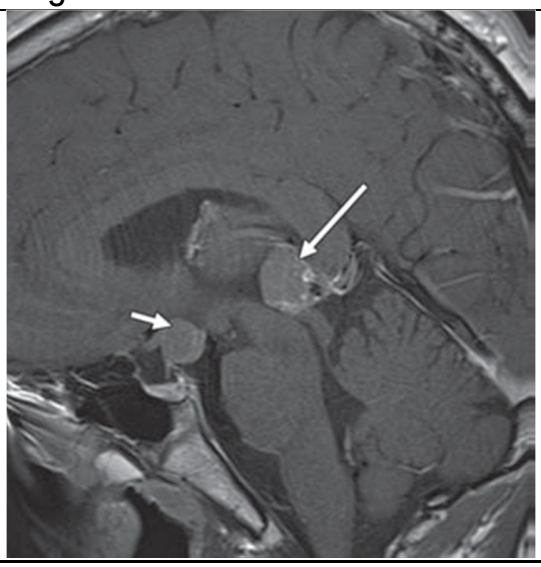
- Clinical: Typically present with *Diabetes Insipidus*

Location:

- 60% Pineal
- 35% Suprasellar

Imaging findings:

- T1: Low signal
- T2: High signal
- +C: Heterogeneous enhancement

Sagittal T2	Sagittal T1+C
	

4) Hypothalamic Hamartoma

- Heterotopic gray matter generally located in tuber cinereum

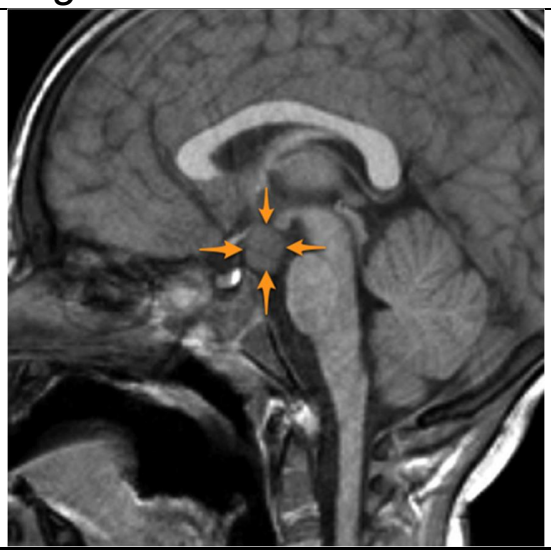
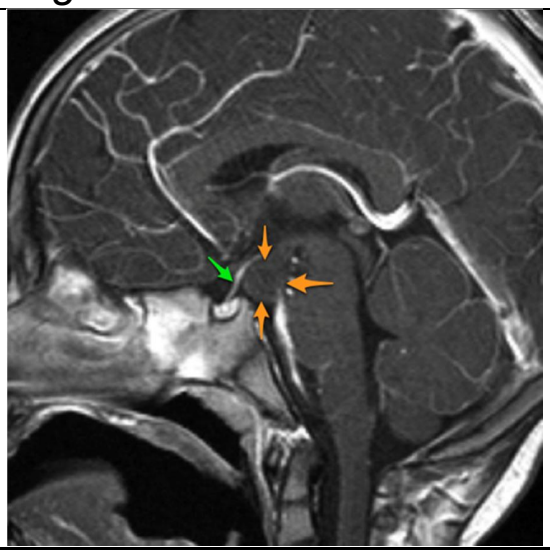
- Age: 1-3yrs
- Sex: M=F

Clinical picture:

- Large lesions cause gelastic seizures
- Small lesions have *precocious puberty*

Imaging findings:

- Sessile or pedunculated lesion
- Attached to the tuber cinereum
- Between the pituitary stalk and mamillary body.
- T1: Iso
- T2: Iso
- +C: *No enhancement*

Sagittal T1	*Sagittal T1+C*
	

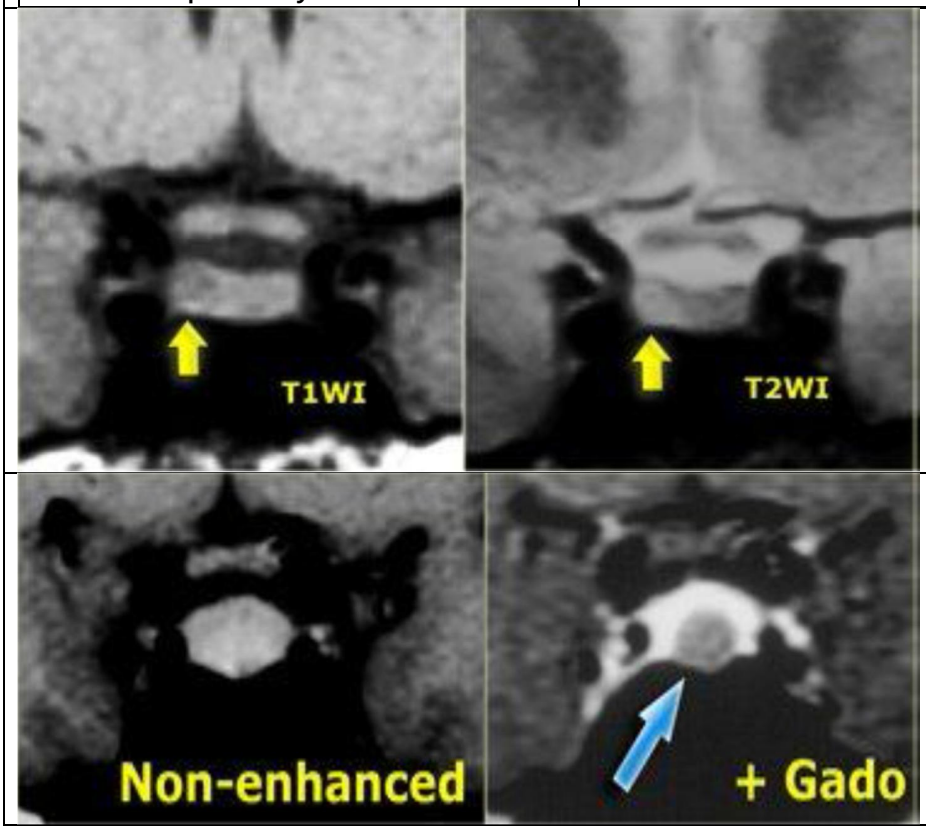
ii- Adult Sellar and Suprasellar Lesions				
	Pitutary Microadenoma	Pitutary Macroadenoma	Meningioma	Craniopharyngioma
Incid		(1st Common)	(2nd common)	2 peaks middle age
Site	Intra sellar	Predominant intra sellar with supra sellar extension	1)Sphenoid ridge 2)diaphragma sella 3)Tuberculum sella	Predominant supra sellar with smaller intra-sellar component
Rad	< 1 cm	> 1 cm - *Snowman = Figure of 8* - Cyst formation common - No calcification - Hge common	- Extra axial - Lobulated - No cystic component - Gross calcification - No hge	- Rounded - No cystic component - No calcification
CT	Hypo / Iso dense	Hetrogenous hyperdense	Homogenous hyperdense	Heterogeneous
MRI	T1 : Hypo T2 : Hyper	T1 : Hypo T2 : Hyper	T1 : *Iso* T2 : *Iso*	Low T1 high T2
+C	Enhance less rapidly than normal pituitary	Heterogeneous intense enhancement	Strong homogenous. enhance. with dural tail sign	No enhancement

1) Pituitary Microadenoma

- < 10 mm in diameter
- Located within the pituitary gland.
- Ipsilateral convex upper bulge
- Contralateral stalk deviation
- Sella floor thin
- Dynamic MRI: adenoma enhances slower than normal gland

- Prolactinoma 30%
- Null cell 25%
- GH 20%
- ACTH 10%
- FSH/LH 10%
- PRL-GH 5%
- Mixed, TSH 1%-5%

T1: slightly hypointense compared to normal pituitary tissue	T2: slightly hyperintense.



2) Pituitary Macroadenoma

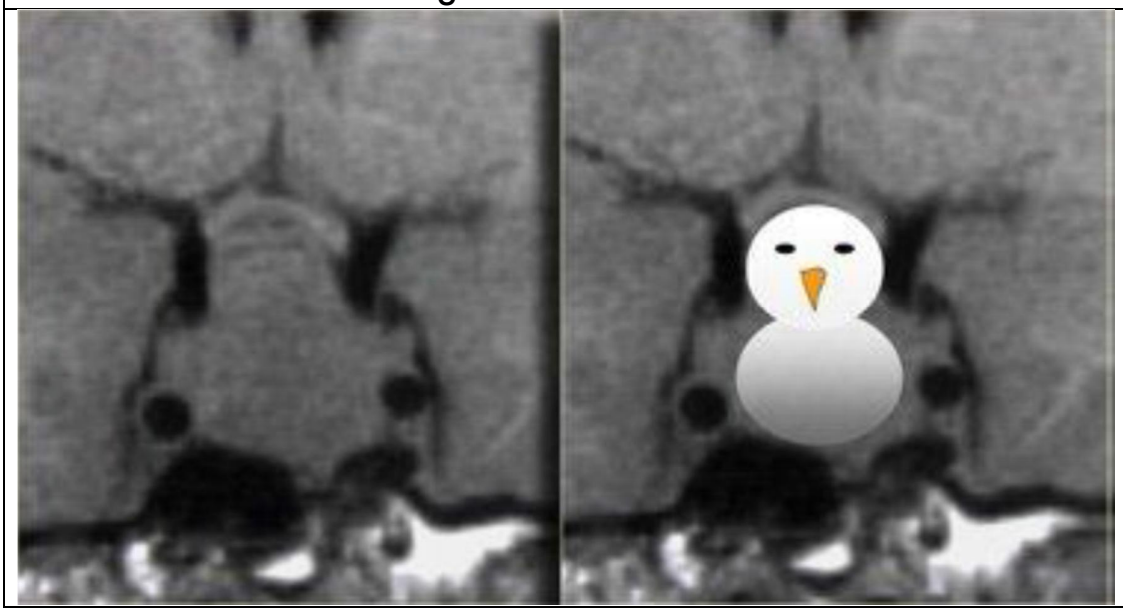
WHO Grade 1

- Intra-sellar tumors => Expand the sella turcica
- Intra-sellar & Suprasellar soft tumors => indent at the diaphragma sellae => *'Snowman' configuration.*
- Supra-sellar extension => elevation and compression of the optic chiasm.
- Para-sellar extension => Invasion of cavernous sinus and ICA encasement => Unresectable
- Trans-sphenoidal extension => Nasopharyngeal mass

Imaging findings:

- T1: Low
- T2: High
- +C: heterogeneous enhancement

❖ 'Snowman' configuration



T1: Low signal	+C: Sellar and suprasellar heterogeneously enhancing lesion
Parasellar extension => invasion of cavernous sinus + ICA encasement	*Trans-Sphenoidal Extension => Nasopharyngeal Mass*

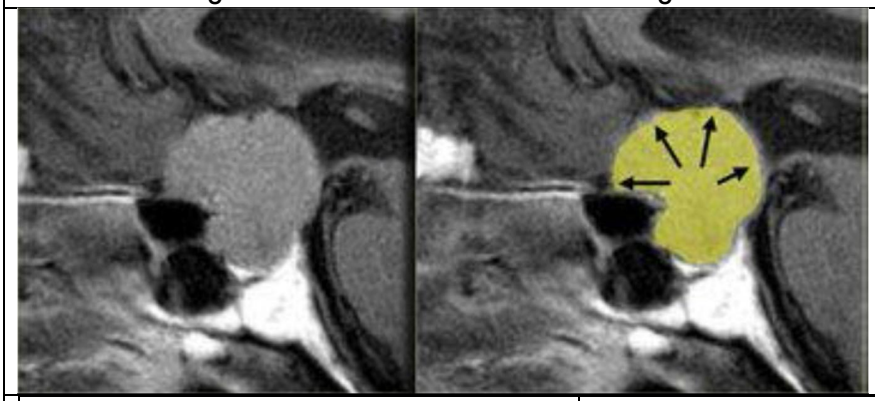
3) Suprasellar / planum sphenoidale Meningioma

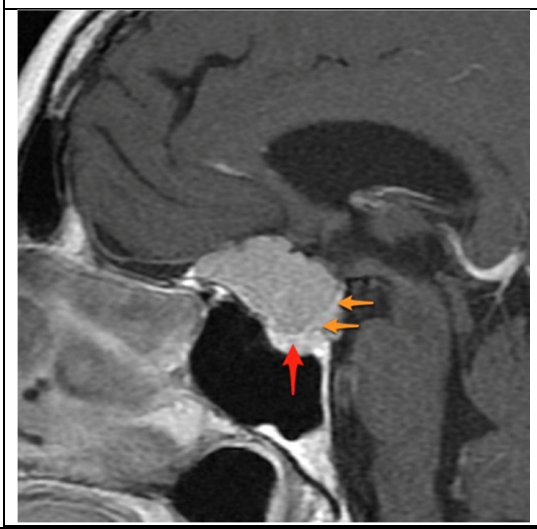
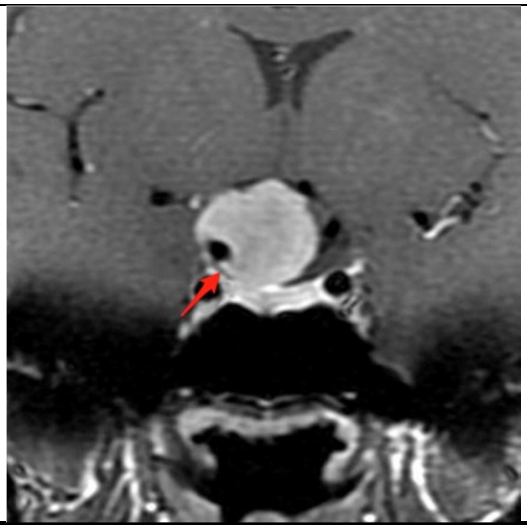
- Most common primary intracranial tumor in adults.

- Age: Middle age
- Sex: F>M

Imaging findings:

- Extra-axial suprasellar / planum sphenoidale meningeal based lesion
- T1: iso – Low
- T2: iso – High
- +C: homogenous enhancement + Meningeal tail



Compression of pituitary Gland + Posterior displacement of pituitary stalk	Suprasellar Mass => Encasement of supraclinoid segment of ICA
	

iii- Infundibulum Lesions

- Granulomatous hypophysitis:
  - TB
  - Sarcoidosis
  - Wegner Granulomatosis
  - Langerhans Cell Histiocytosis (Eosinophilic Granuloma)
- Meningeal Carcinomatosis:
  - Metastasis
  - Lymphoma
  - Leukemia
- Lymphocytic Hypophysitis

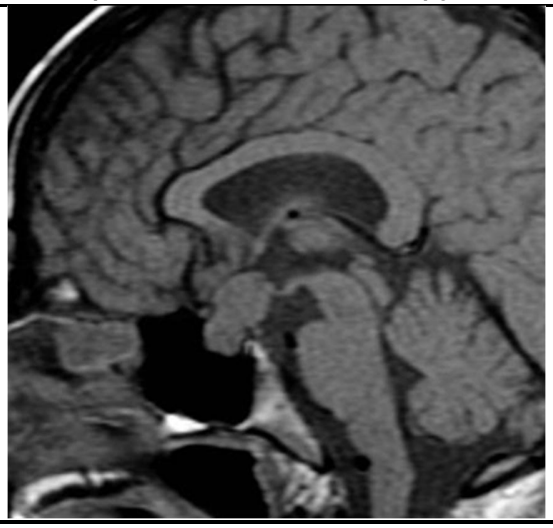
❖ Lymphocytic Adeno- Hypophysitis

- Lymphocytic hypophysitis (LH) is uncommon non-neoplastic inflammatory condition that affects the pituitary gland.

- It is closely related to other inflammatory conditions in the region:
  - ➢ Orbital Pseudotumour
  - ➢ Tolosa-Hunt syndrome

Clinical Picture:

- Occurs during late pregnancy or shortly after delivery
- F >>> M
- Presents with pituitary insufficiency & visual changes
- Amenorrhea or inability to lactate
- Diffuse enlargement of gland, stalk or both.
- May mimic pituitary hyperplasia or adenoma
- Responds to steroid therapy.



iv- Cystic Sellar / Suprasellar Lesions

- Rathke-Cleft cyst
- Pituitary Apoplexy

- Cystic pituitary macroadenoma
- Cystic craniopharyngioma

- Arachnoid cyst
- Epidermoid
- Dermoid
- Lipoma
- Teratoma

1) Rathke-Cleft cyst

- Non-neoplastic, epithelium-lined cysts arising from the embryologic remnants of Rathke's pouch in the pituitary gland.

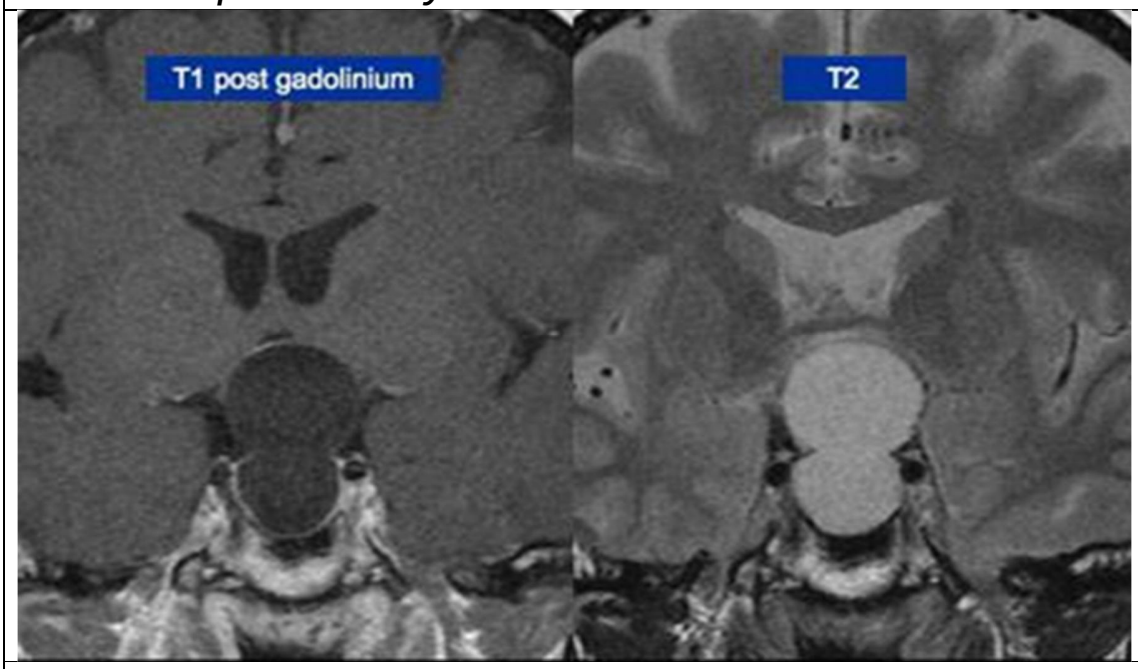
Locations:

- 40% intrasellar (between anterior and intermediate lobes of pituitary).
- 60% Intrasellar + suprasellar extension.
- Rare, Purely suprasellar location.

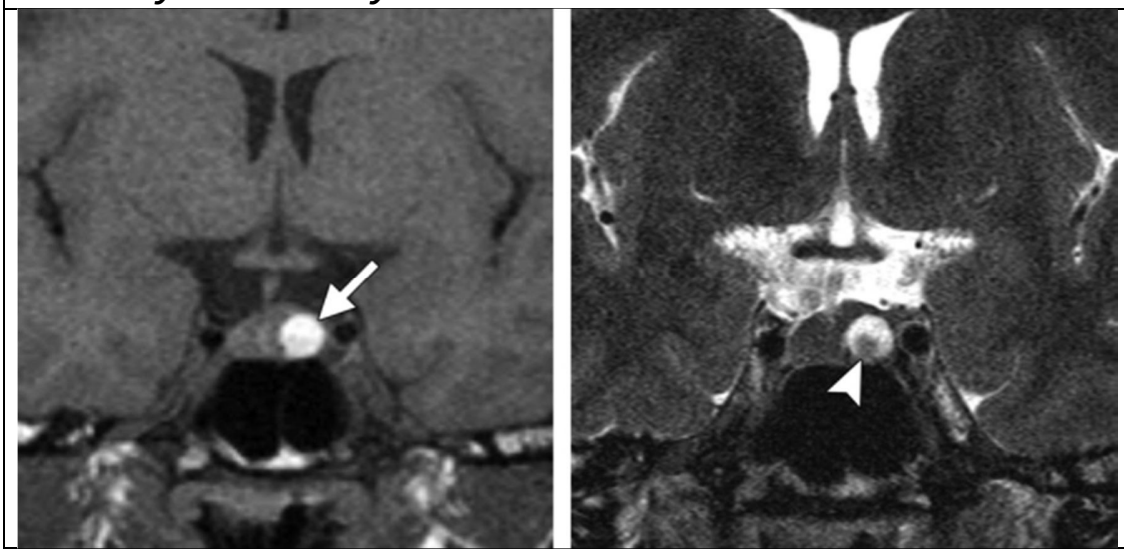
Imaging Findings:

- Well defined midline cystic lesion
- T1
    - 50% are hyperintense (high protein content)
    - 50% are hypointense
- T2
    - 70% are hyperintense
    - 30% are iso or hypointense
- T1 C+ (Gd)
    - No contrast enhancement of the cyst is seen,
    - Thin enhancing rim of surrounding compressed pituitary tissue may be apparent
- In 70 - 80% of cases a small non-enhancing intracystic nodule can be identified which is virtually pathognomic of a Rathke's cleft cyst.
    - T1: Hyper intense to surrounding fluid
    - T2: Hypointense to surrounding fluid
- Occasionally, a fluid-fluid level may be seen (particularly if there has been a haemorrhage)

Sellar + Suprasellar Cyst



Sellar cyst + Intracystic nodule



2) Pituitary Apoplexy

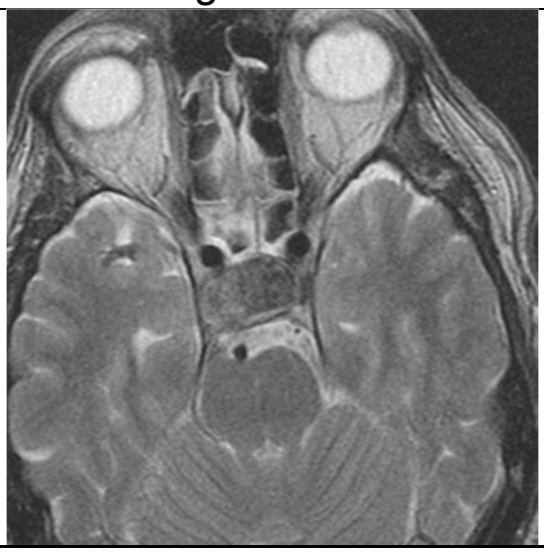
- is an acute clinical syndrome caused by either haemorrhage or infarction of the pituitary gland

Clinical presentation:

- Sudden headache
- Loss of visual acuity with a chiasmal field defect
- Oculomotor palsies

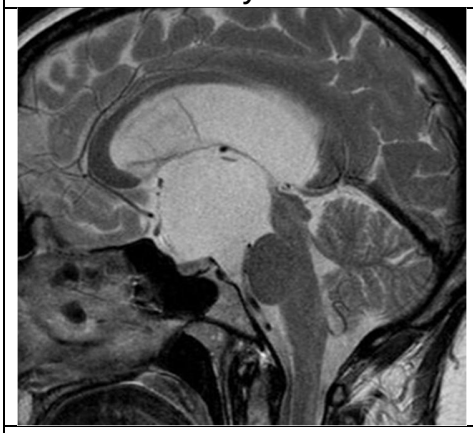
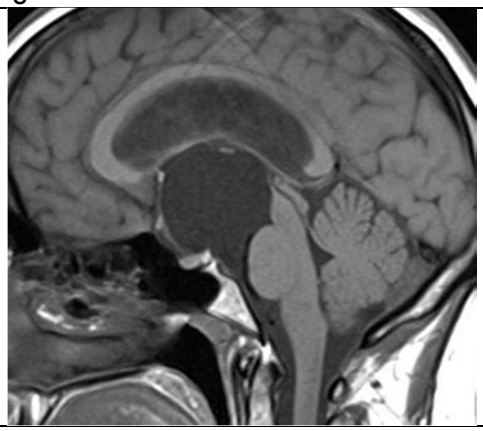
Predisposing Factors

- Medical treatment of a prolactinoma (especially with bromocriptine)
- Prior irradiation of the mass
- Postpartum (Sheehan syndrome)
- Cerebral angiography
- Trauma and surgery
- Anticoagulation
- Changes in intracranial pressure

T1: High signal	*T2: Low signal*
	

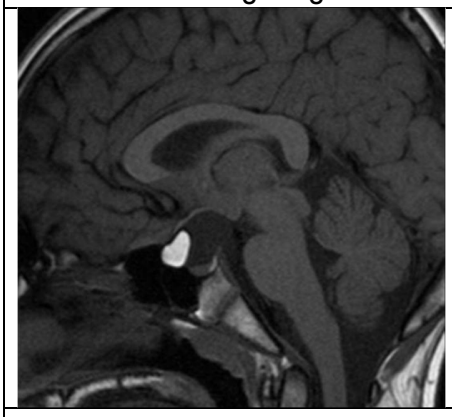
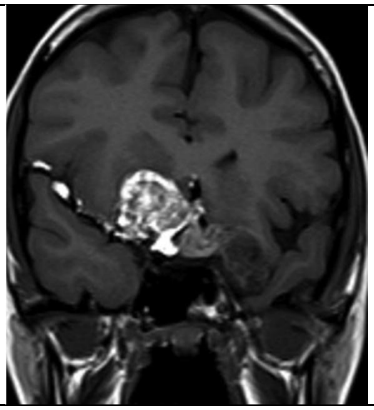
3) Suprasellar Arachnoid Cyst

- Extra-axial cystic lesion of CSF like signal

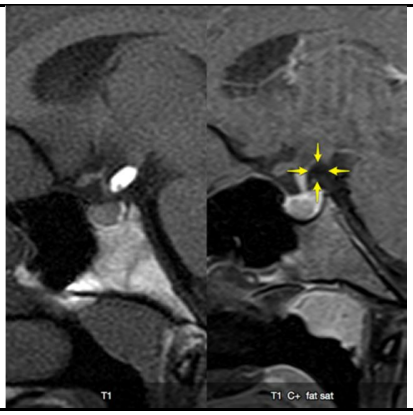



4) Suprasellar Teratoma

- Mixed fluid + Fat + calcification
- T1: Low + high signal

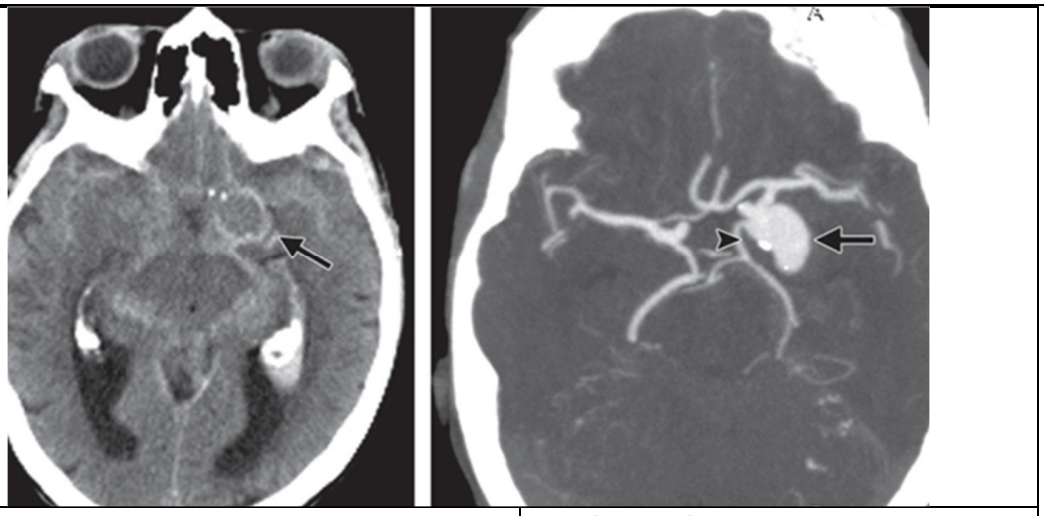
5) Suprasellar Lipoma



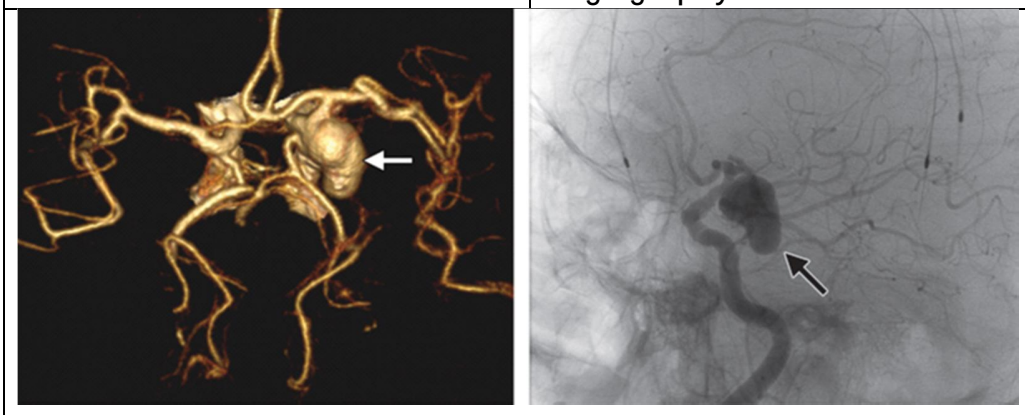
v- Vascular Lesions

Suprasellar Aneurysm

- 90-95% arise from Circle Of Willis
    - Anterior circulation: 90%
        - ACA / Acom complex: ~30 - 40%
        - ICA / Pcom junction: ~30%
        - MCA (M1/M2 junction) bifurcation: ~ 20 - 30%
    - Posterior circulation: 10%
        - Basilar tip
        - SCA
        - PICA



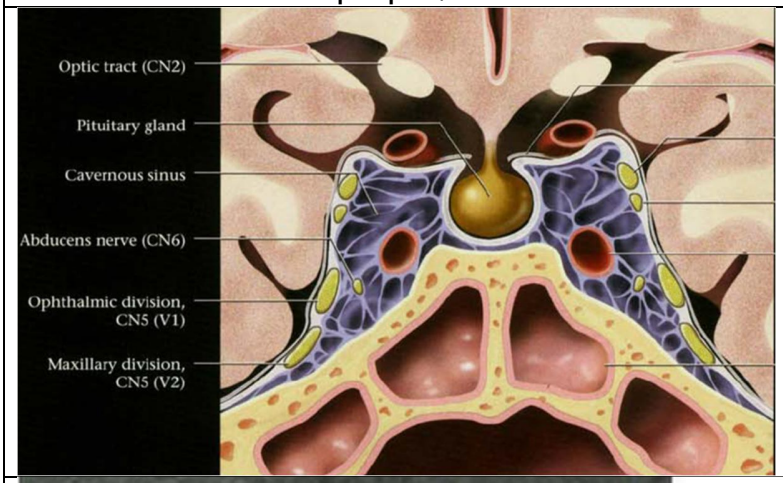
3D CTA | Angiography

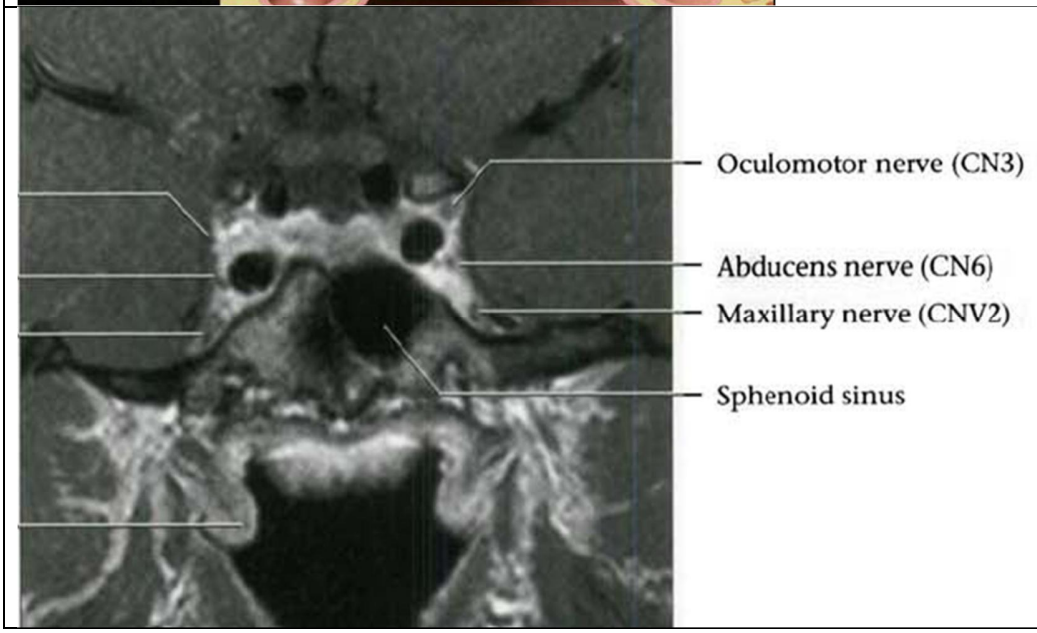


B- Parasellar / Cavernous Sinus Lesions

Contents:

- CN3 lies within superior lateral dural wall
- CN4 just below CN3
- V1 (ophthalmic division of CN5) in lateral wall below CN4
- V2 (maxillary division of CN5) is most inferior CN in lateral wall
- V3 (mandibular division of CN5) does NOT enter Cavernous sinus proper (passes from Meckel cave inferiorly into foramen ovale)
- CN6 lies within CS proper, next to ICA





- ❖ Congenital / Developmental :
  - o Arachnoid cyst
  - o Dermoid
  - o Epidermoid
- ❖ Vascular :
  - o Aneurysm
  - o Carotid – cavernous fistula
  - o Cavernous sinus thrombosis
  - o Cavernous hemangioma
- ❖ Infectious / Inflammatory:
  - o Tolosa-Hunt syndrome
  - o TB / Sarcoidosis / Wegener Granulomatosis
  - o Fungal
- ❖ Meninges :
  - o Meningioma
- ❖ Cranial nerves :
  - o Peripheral nerve sheath tumor: Schwannoma – Neurofibroma
  - o Malignant peripheral nerve sheath tumor
  - o Perineural spread
- ❖ Sphenoid sinus :
  - o Sinusitis – Fungal
  - o Mucocele
- ❖ Base of Skull :
  - o 1ry : Chordoma - Chondrosarcoma
  - o 2ry : Metastasis
- ❖ Nasopharyngeal => parasellar extension:
  - o Nasopharyngeal carcinoma
  - o Nasopharyngeal angiofibroma
  - o Nasopharyngeal RMS
- ❖ Sellar / Suprasellar => Parasellar extension:
  - o Pituitary macroadenoma

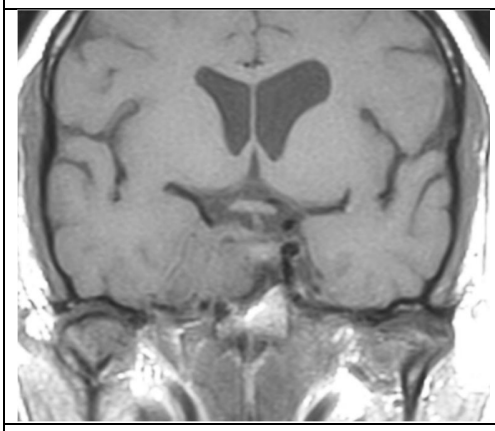
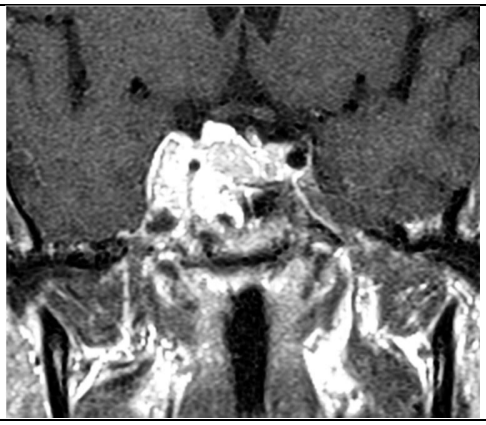
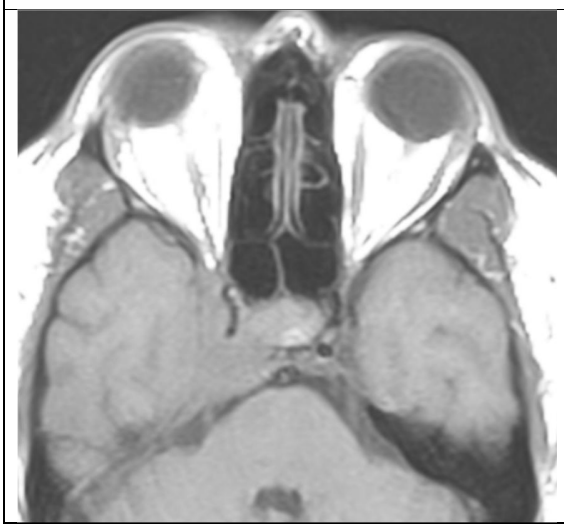
i) Benign Parasellar Lesions		
	Schwannoma	Meningioma
Incidence	6-8 %	20%
Age	40-60 Y	40 – 60 Y
Imaging findings	-Extra axial -heterogeneous hypodense	-extra axial -homogeneous hyper dense
Ca	Rare	25 %
Cystic	Common	Rare
MRI	T1 : Hypo T2 : Hyper	T1 : Iso T2 : Iso
+C	Strong heterogeneous enhancement	Strong Homogenous enhancement dural base & tail

1) Parasellar Meningioma

- Meningioma is most common extraaxial lesion in adults
- Age: Middle age
- Sex: F > M

<u>Imaging findings:</u>

- Extra-axial meningeal based lesion
- Homogenous density / signal
- CT: hyperdense
- T1: Iso – Low
- T2: Iso – High
- +C: Homogenous enhancement + dural tail

Coronal T1	Coronal T1+C
	
Axial T1	Axial T1 +C
	

2) Parasellar Schwannoma

- Schwannoma is most common tumor of peripheral nerves
- Age: Middle age
- Sex: F= M

Intracranial Location:

- Vestibulocochlear (Acoustic / CPA) (1st common)
- Trigeminal schwannoma (2nd common)
- Facial schwannoma (3rd common)

Imaging findings:

- Well defined extra-axial lesion
- Dumbbell shaped lesion:
  - Preganglionic (cisternal) confined to CPA
  - Ganglionic (gasserian ganglion) confined to Meckel's cave
  - Postganglionic either confined to the cavernous sinus
- Cystic degeneration : common
- Calcification: rare
- T1: Low
- T2: High signal
- +C:
  - Small tumours => Homogeneous enhancement
  - Larger tumours => heterogeneous enhancement

Axial T2	Axial T1+C

ii) Destructive Parasellar Lesions

1) Parasellar Chordoma

- Rare intracranial tumor
- Age: Adults
- Sex: M > F

Location:

- Sacrococcygeal: 30-50%
- Spheno-occipital: 30-35%
- Vertebral body: 15-30%

Imaging findings:

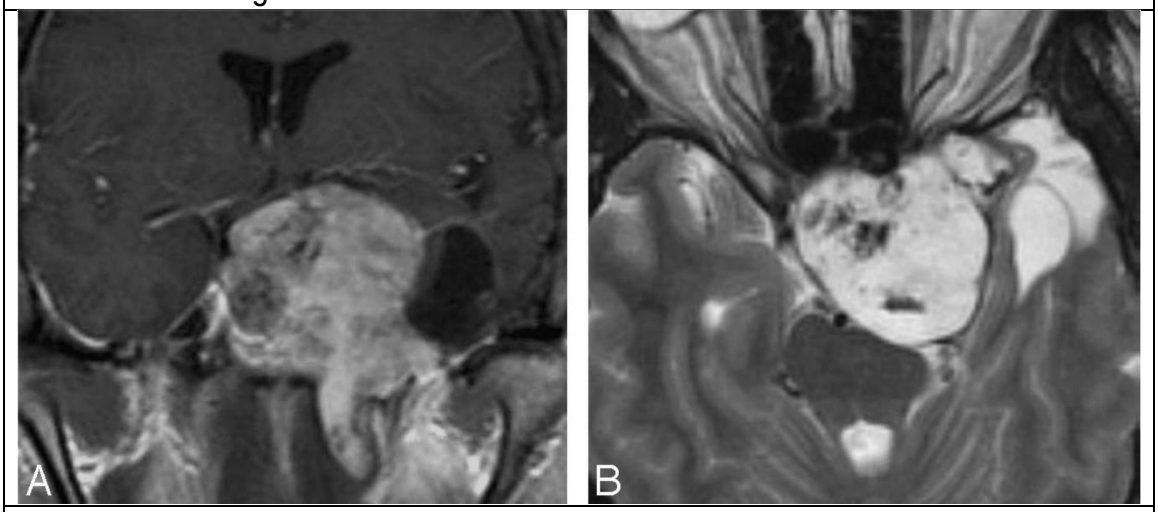
- Midline primary clival tumor (spheno-occipital synchondrosis)
- Well defined destructive lesion
- Cystic degeneration / hemorrhage / calcification: common
- T1: low signal
- T2: High signal
- +C: heterogeneous enhancement

Axial T1	Axial T1+C

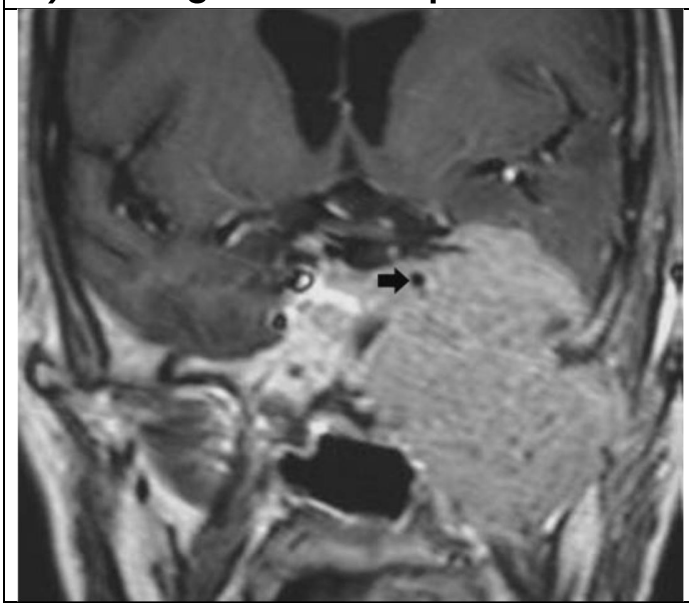
2) Parasellar Chondrosarcoma

Imaging findings:

- Originate from the petroclival synchondrosis
- Infiltrate destructive lesion => Petrous apex + Cavernous sinus
- CT: Chondroid matrix = amorphous calcification: Rings and arcs
- T1: Low signal
- T2: Characteristic high signal
- +C: Hetrogenous moderate / marked enhancement



3) Malignant Peripheral Nerve Sheath Tumor

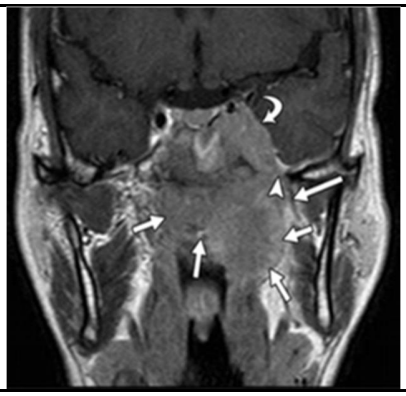


4) Nasopharyngeal Carcinoma

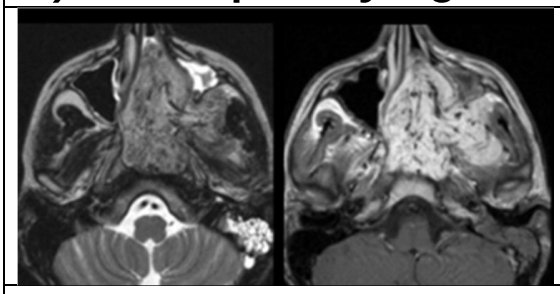
- Nasopharyngeal carcinoma is the most common primary malignant extracranial neoplasm to invade the cavernous sinus.

<u>Intracranial extension via</u>

- Skull base erosion
- Perineural spread along branches of the trigeminal nerve
- Petro-occipital synchondrosis
- Foramen lacerum
- Carotid canal



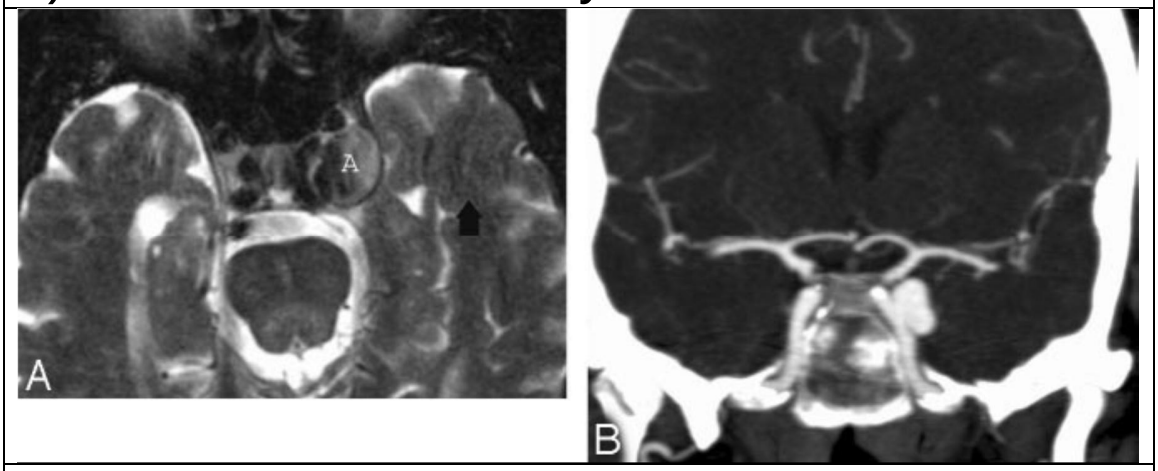
5) Nasopharyngeal Angiofibroma



6) Nasopharyngeal RMS

iii- Vascular Lesions:

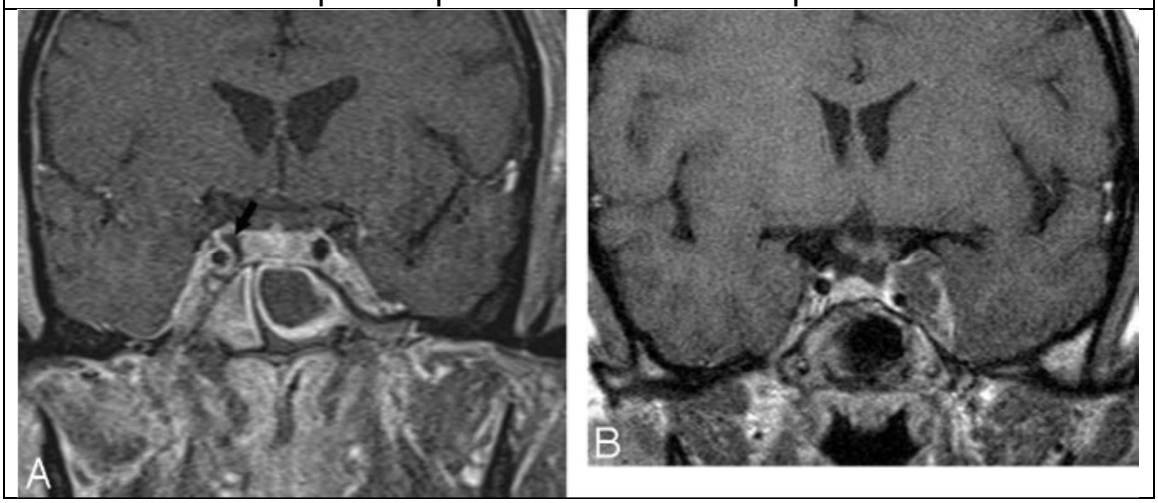
1) ICA Cavernous Aneurysm



2) Cavernous sinus Thrombosis

- CS thrombosis may be secondary to infection of the sinonasal cavities, orbits, and/or the middle third of the face.

- Changes in signal intensity and/or in the size and contour of the CS.
- Enhancement of the peripheral margins of an enlarged CS.
- Dilation of the superior ophthalmic veins and exophthalmos



3) Carotid Cavernous Fistula

Direct = High Flow shunt:

Type A: Direct ICA-cavernous sinus (not true dAVF)

Indirect = Dural = Low flow shunt:

Type B: ICA dural branch - shunt to cavernous sinus

Type C: ECA dural branch - shunt to cavernous sinus

Type D: ECA/ICA dural branches - shunt to cavernous sinus

Imaging findings:

- Multiple signal-intensity void in the cavernous sinus
- MRA: high flow signal in the cavernous sinus
- Enlarged superior ophthalmic vein
- Proptosis

MRA: High flow signal in the left cavernous sinus (arrow)	T2: Multiple flow voids in the left cavernous sinus (arrows).

iv- Infectious / Inflammatory Lesions

1) Tolosa-Hunt Syndrome

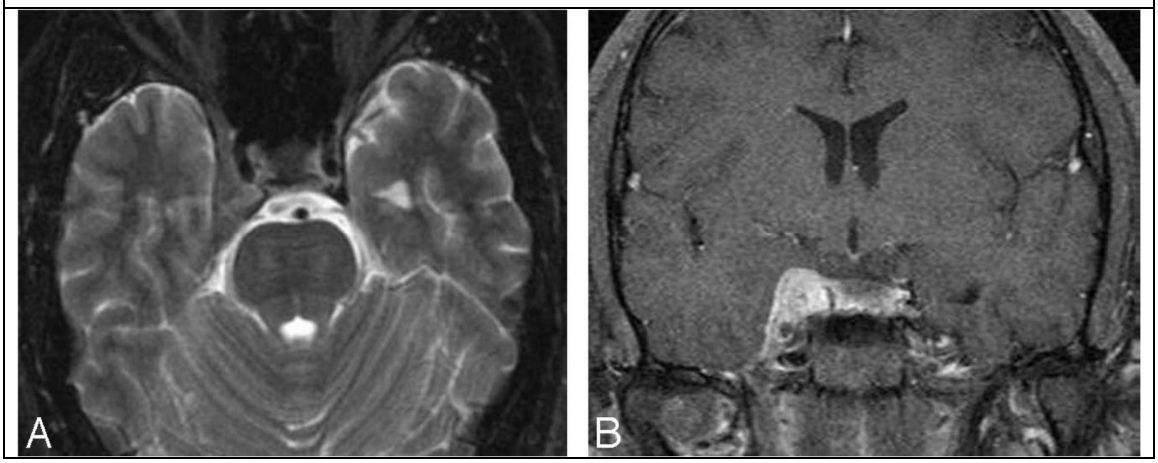
- Tolosa-Hunt syndrome is a term applied to a retro-orbital pseudotumor extending to the CS.
- Low-grade nonspecific inflammatory process with lymphocytes and plasma cells.

Its clinical triad :

- Unilateral ophthalmoplegia
- Cranial nerve palsies
- Dramatic response to systemic corticosteroids

Imaging Findings:

- Enlargement of the cavernous sinus unilaterally
- T1: iso – Low signal
- T2: Low / High signal
- +C: enhancement



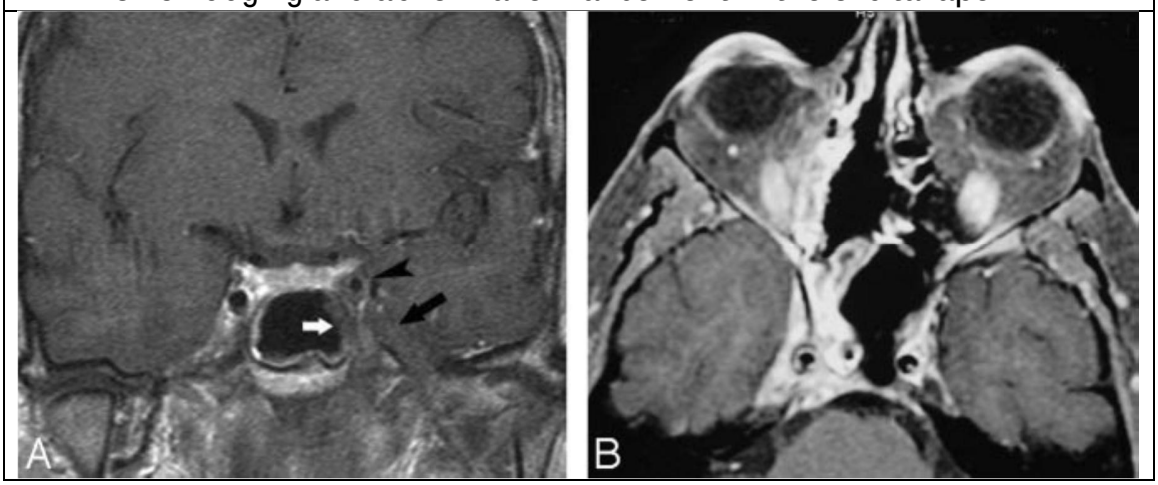
2) Fungal Infection

- Invasive aspergillosis may affect the sphenoid sinus in immunocompromised patients

- Rhinocerebral mucormycosis is acute fulminant opportunistic infection, which may affect the orbits and paranasal sinuses

Imaging Findings:

- Cavernous sinus invasion:
  - T1 & T2: low signal intensity, which is attributed to the presence of ferromagnetic elements and calcium in the fungal and mucous concretions.
  - +C: It exhibits intense inhomogeneous contrast enhancement
  - Thrombosis and thickening of the ICA walls
  - Smudging and abnormal enhancement in the orbital apex



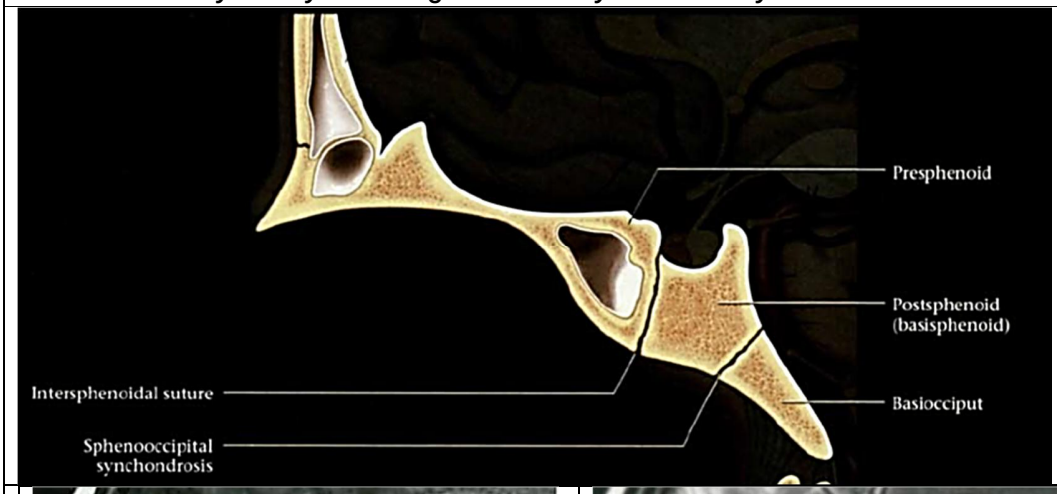
C. Basisphenoid / Clival / Foramen magnum

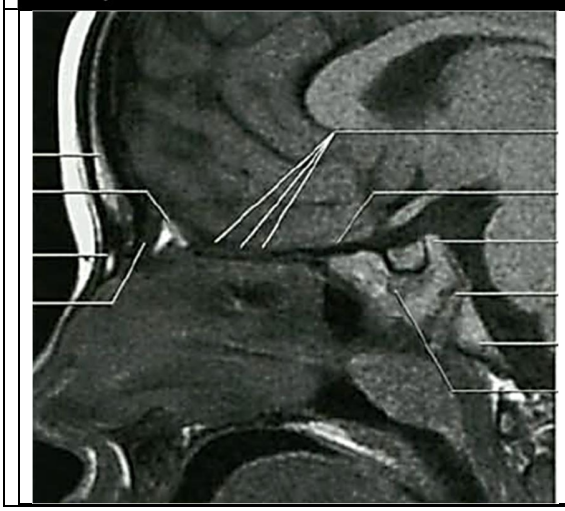
Clivus:

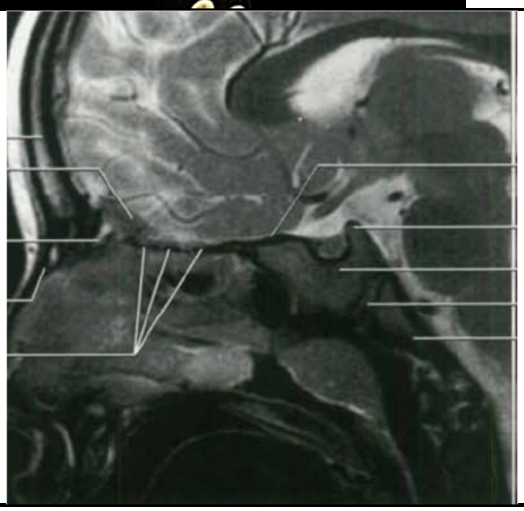
- Fusion of basisphenoid and basiocciput fuse
- < 25y: Low T1 signal = red marrow
- > 25 y: High T1 signal = fatty marrow

Sphenooccipital synchondrosis

- Between basisphenoid and basiocciput
- Open until 14 years
- fuses by - 16 years in girls & - 18 years in boys



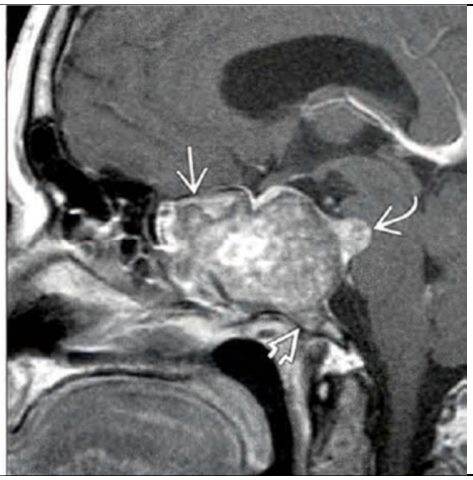




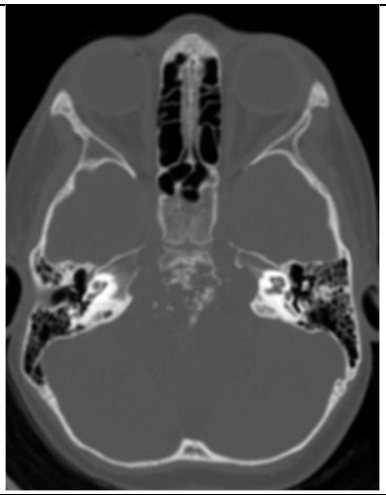
DD:
• Chordoma • Chondrosarcoma • Meningioma • Pituitary macroadenoma • Nasopharyngeal carcinoma • Metastasis • Plasmacytoma • Ecchordosis Physaliphora

	Chordoma	Chondrosarcoma
	Rare clival malignant tumor Arising from remnants of cranial end of primitive notochord.	Rare skull base tumor Arise from embryonal rest cells
Location	•Sacrococcygeal: 30-50% •Spheno-occipital: 30-35% •Vertebral body: 15-30%	Petro-occipital fissure (2/3) Anterior basisphenoid (1/3)
	Midline	Paramidline
	Spheno-occipital synchondrosis.	Petroclival synchondrosis
	> 30 y M > F	
CT	Bone destruction Bone fragments Cystic degeneration / hemorrhage / calcification	Bone destruction Chondroid matrix = Amorphous calcification: Rings and arcs
MRI	T1: Low signal T2: Characteristic High signal + Low signal foci of calcification	
+C	Heterogeneous Enhancement	
ADC	Low < 1.8	High > 1.8

1) Clival Chordoma



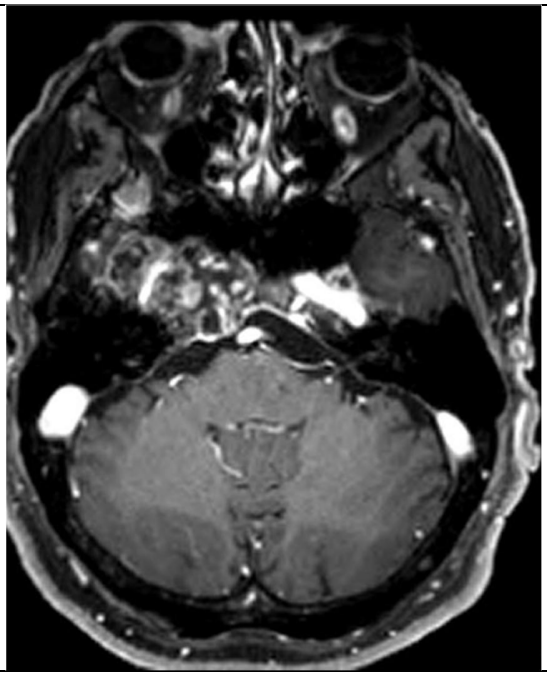
Intratumoral Calcifications



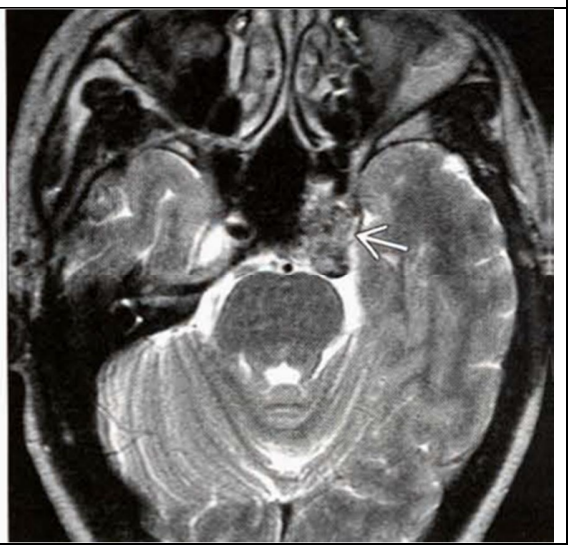
Clival chordoma => Anterior growth

2) Chondrosarcoma

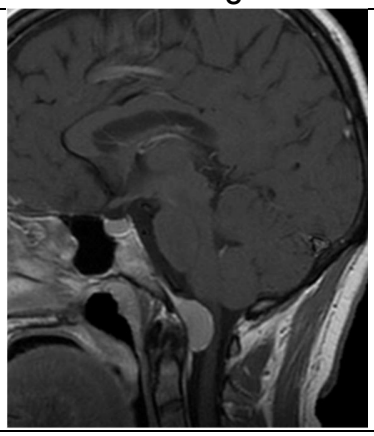
- Petro-occipital fissure => Petrous apex + Clivus / Basisphenoid
- Anterior growth => Parasellar / cavernous sinus region
- Posterior growth => prepontine + CPA region

Axial T2	Axial T1+C FS
	

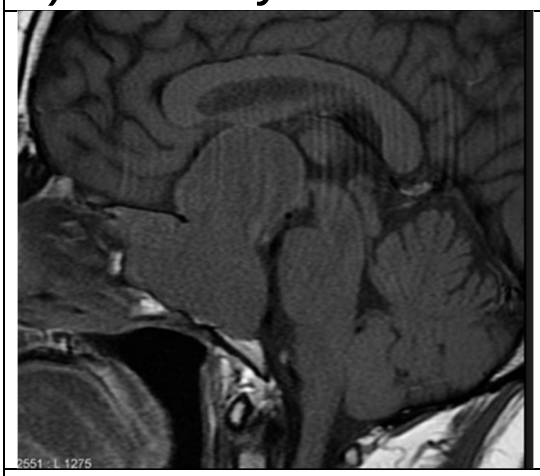
Intratumoral calcification

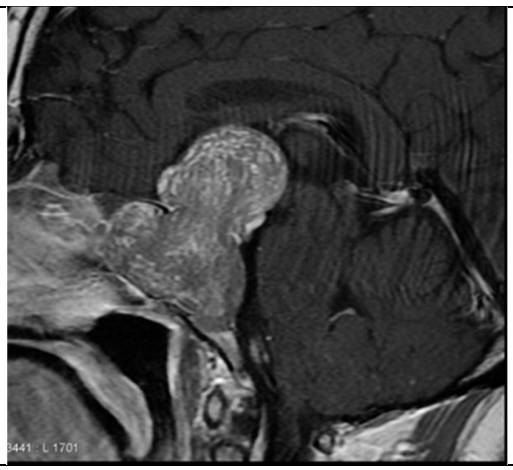


3) Meningioma

Clival meningioma	Foramen magnum meningioma
	

4) Pituitary Macroadenoma





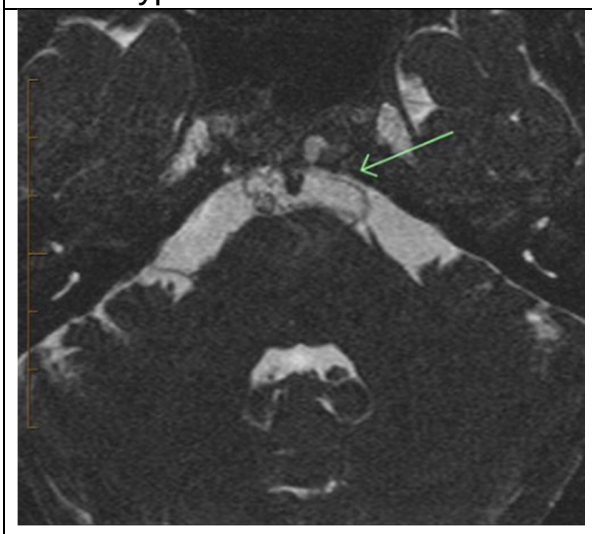
5) Nasopharyngeal Carcinoma

6) Ecchordosis Physaliphora

- Congenital benign hamartomatous lesion derived from notochord remnants
- Location: Retroclival prepontine region

<u>Radiology Findings:</u>

- Osseous retroclival stalk
- No clival bony destruction
- No enhancement.
- T1: hypointense
- T2: hyperintense



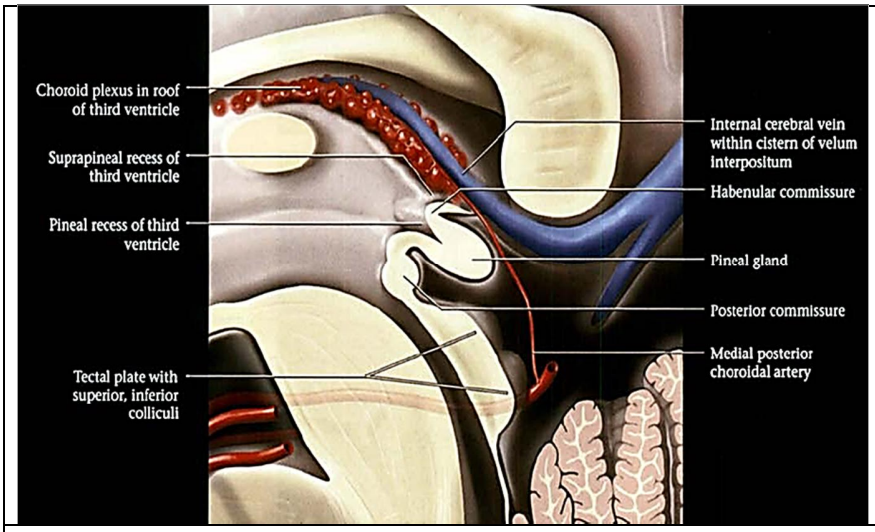
D- Pineal / Quadrigeminal Cistern Lesions

❖ Pineal gland:

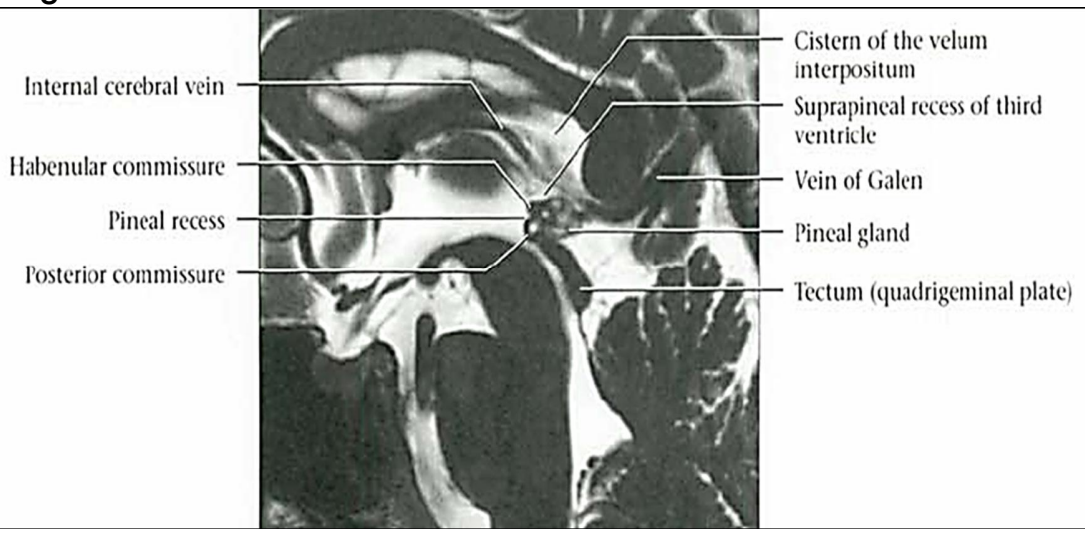
- is a small reddish-brown structure that derives its name from its pinecone-like shape.
- Size from 10 to 14 mm;
- Midline
- Above the tectal plate and superior colliculi
- Below the splenium of the corpus callosum and the vein of Galen
- is attached to the superior aspect of the posterior border of the 3rd ventricle
- The pineal gland secretes melatonin, which is involved in diurnal rhythms.
- The pineal gland does not have a blood-brain barrier and therefore enhances on contrast material–enhanced images

❖ Quadrigeminal Cistern

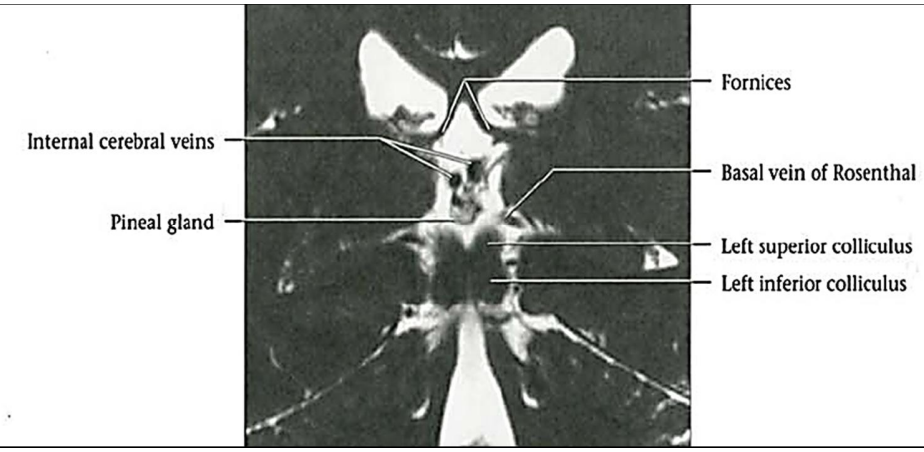
- Pineal gland
- Trochlear nerves (CN4)
- P3 PCA segments
- Medial & lateral posterior choroidal arteries
- Vein of Galen (VofG) + tributaries


Choroid plexus in roof of third ventricle
Suprapineal recess of third ventricle
Pineal recess of third ventricle
Tectal plate with superior, inferior colliculi
Internal cerebral vein within cistern of velum interpositum
Habenular commissure
Pineal gland
Posterior commissure
Medial posterior choroidal artery

Sagittal T2



Coronal T2



❖ Origin Of Pineal Lesions

- Pineal parenchyma (14%–27%)
  - Low-grade pineocytoma.
  - Intermediate-grade pineal parenchymal tumor of intermediate differentiation (PPTID)
  - Highly malignant pineoblastoma
- Germ cell neoplasms (40%)
  - Germinoma
  - Non-Germinoma
    - Teratoma
    - Yolk sac tumor
    - Embryonal cell carcinoma
    - Choriocarcinoma
- Metastasis.
- Tectal plate:
  - Glioma
- Vein of Galen:
  - Aneurysm
  - Thrombosis
- Splenium of corpus callosum:
  - Glioblastoma multiform (GBM)
  - Lymphoma
  - Tumefactive MS
- Congenital Quadrigeminal / supracerebellar cistern:
  - Lipoma / Dermoid / Arachnoid / Epidermoid

❖ Clinical Picture 3P:

- Parinaud syndrome: consists of a failure of conjugate vertical eye movement, mydriasis, failed ocular convergence, and blepharospasm due to compression or invasion of the tectal plate.
- Precocious puberty may be seen with germ cell tumors (GCTs).
- Pressure effect on the aqueduct of Sylvius => Hydrocephalus => patients may also develop headache, nausea, and vomiting as a result of increased intracranial pressure.
- Diabetes insipidus

❖ Pineal Calcification

- Concentric calcifications (corpora arenacea)
- Appear in adolescence and are seen in 40% of patients 17–29 years old.

i- Pediatric Pineal Lesions

	Pineoblastoma	Germinoma	Teratoma
Inc.	Uncommon	Most common	2nd common
Age	< 20 y	< 20 M	Newborn
Sex	F > M	M > F	M > F
Ass.	*Trilateral retinoblastoma*	Pineal + supra sellar	Increase alpha FP
CT	-Large lobulated -Heterogeneous hyper dense *-Exploded peripheral calcification*	-Well circumscribed -Homogenous hyper dense *-Central calcification*	- Lobulated - Fat density - Calcification - Cystic component
MRI	-T1:- hypo -T2 :- hyper	**Dense cell packing:* - T1:- Iso / hyper - T2:- Iso / hyper	T1:- Fat :- hyper Ca:- hypo T2: hetero hyper
+C	Moderate / Marked Heterogeneous enhancement		No / minimal enhancement
Spread	CNS dissemination		No

❖ Germ Cell Tumors

- GCTs arise from residual primordial ectoderm, mesoderm, or endoderm, and each tumor subtype represents the neoplastic correlate of a distinct stage of embryonic development.
- > 50% of the pineal region neoplasms

WHO classification

- Germinoma (1st common)
- Nongerminomatous GCTs.
  - Teratoma (2nd common)
  - Embryonal carcinoma
  - Yolk sac tumor
  - Choriocarcinoma
  - Mixed GCTs.

1) Germinoma

- WHO Grade I
- 1%–2% of all cranial neoplasms

Location:

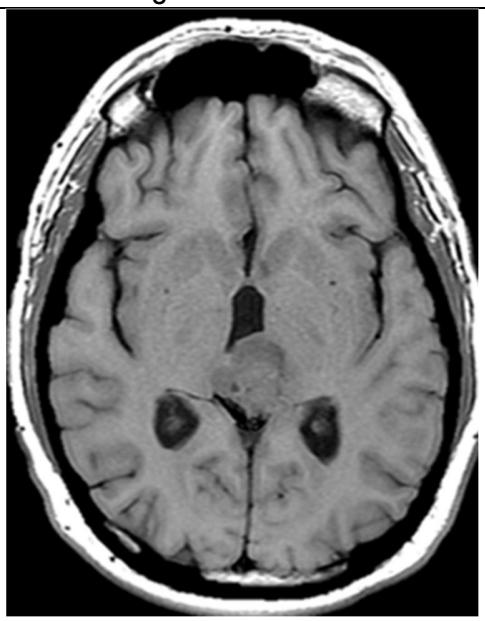
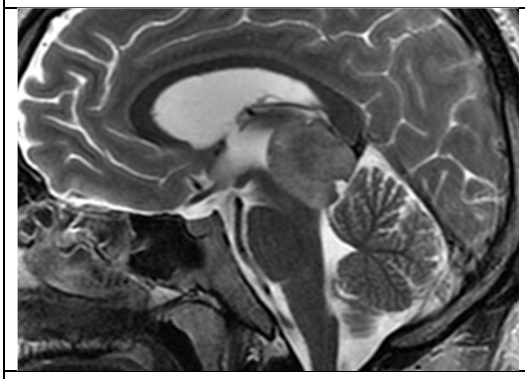
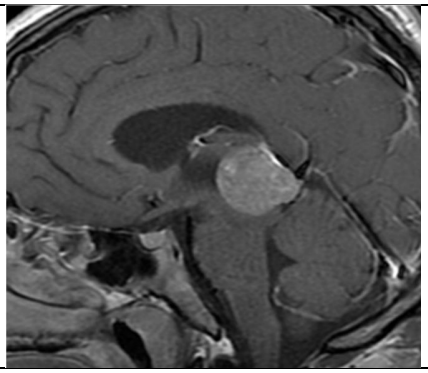
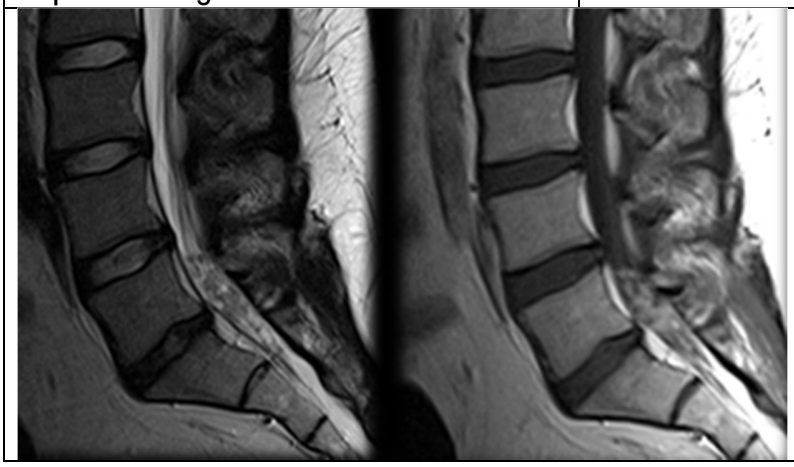
- 65% Pineal region.
- 35% Suprasellar region.

- Age: 10 - 20 years old
- M>F = 10:1

- Prognosis is good (5-year survival at least 90%)
- Radiosenstive

Imaging findings:

- A well defined lesion.
- CT: Hyperattenuating mass: highly cellularity.
- CT: Engulfs the pineal calcifications
- Cystic components
- T1: iso- Low signal
- T2: iso - Low signal
- T1+C: Avid heterogenous enhancement
- DWI: Restricted diffusion
- Hydrocephalus may be present
- Dissemination by CSF.

NCCT: hyperdense mass + Engulf Ca	T1: Low signal
	
T2: Iso - Low	+C: Marked enhancement
	
Leptomeningeal metastasis	
	

2) Teratoma

- Ectodermal, endodermal, and mesodermal lines.

3 Types :

- Mature teratoma (fully differentiated tissue),
- Immature teratoma (complex mixture of fetal-type tissues from all three germ layers and mature tissue elements),
- Teratoma with malignant transformation: the least common form and demonstrates malignant degeneration of the mature tissues.

Location:

- Neonatal:
  - Large Hemispheric Mass
- Juvenile / Young Adult:
  - Pineal or Suprasellar

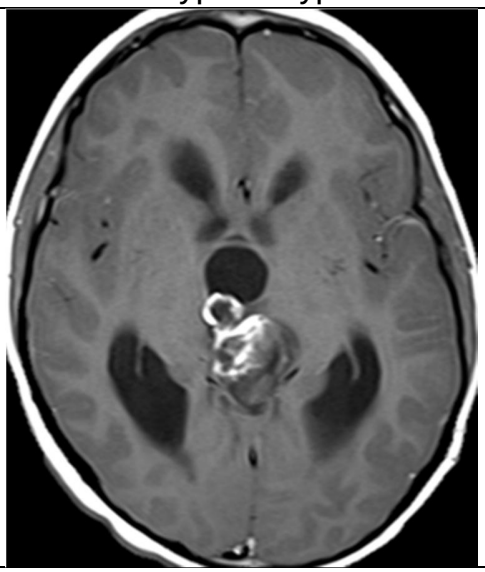
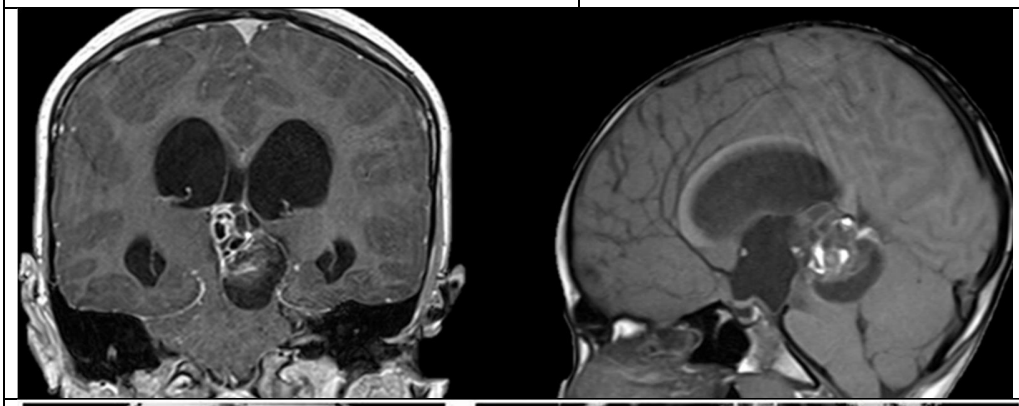
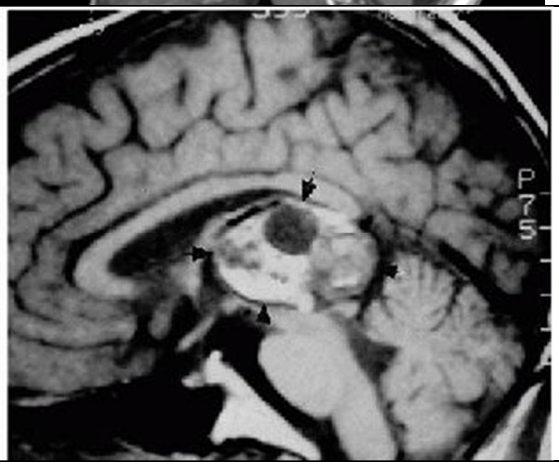
Imaging findings:

CT

- A multiloculated, lobulated lesion with foci of fat attenuation, soft tissue, calcification, and cystic regions.

MRI:

- T1:
  - Hyperintense signal due to fat
  - Variable signal intensity related to calcification.
- T2:
  - Soft-tissue component is iso- to hypointense
  - Cystic component hyperintense.
- T1+C: The soft-tissue component demonstrates enhancement

CT: Fat + fluid density	T1: Mixed hyper + hypo
	
	
	

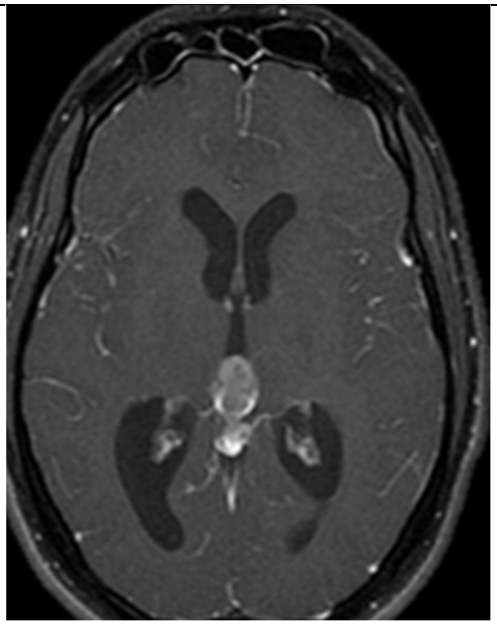
3) Pineoblastoma

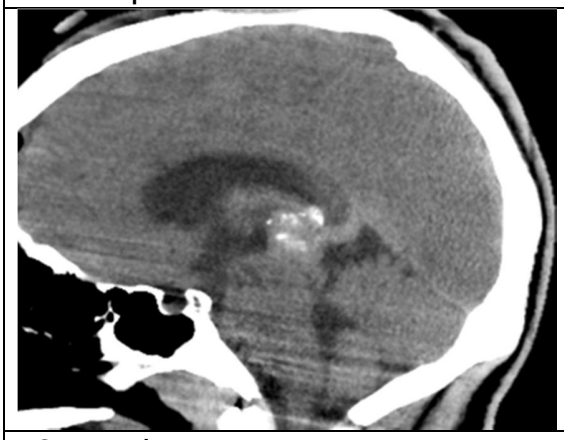
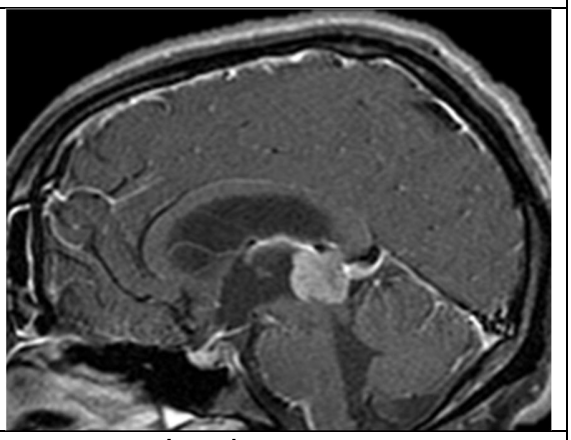
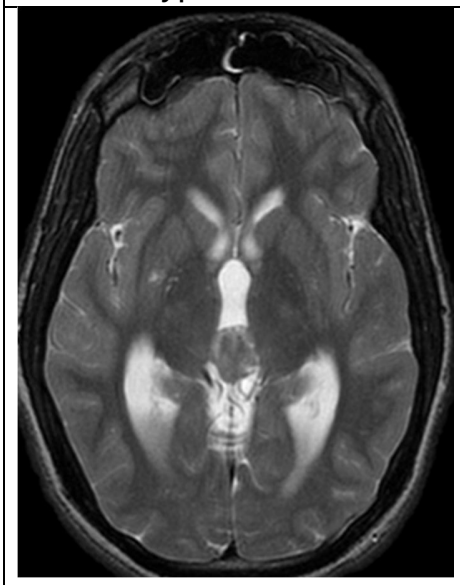
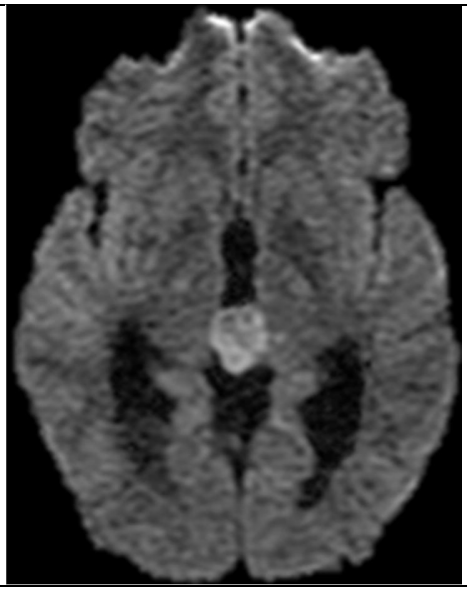
- Highly malignant WHO grade IV
- most primitive form of pineal parenchymal tumors
- 40% of pineal parenchymal tumors
- Embryonal tumors described as a primitive neuroectodermal tumor of the pineal gland
- The 5-year survival is 58%

- Age < 20 y
- Sex = no gender predilection.

Imaging findings:

- A large (typically ≥3 cm), lobulated, typically hyperattenuating mass.
- CT: The pineal calcifications exploded at the periphery of the lesion
- T1: hypo- to isointense
- T2: iso- to mildly hyperintense to the cortex
- T1+C: Heterogeneous enhancement.
- DWI: Restricted diffusion
- Obstructive hydrocephalus.
- CSF dissemination commonly occurs.

T1 : Low signal	+C: Moderate enhancement
	

CT: Peripheral Calcification	+C: Moderate enhancement
	
T2: Iso - hypo	**DWI: Restricted**
	
Leptomeningeal CSF seedling	

5) Tectal Glioma

- Arise from the tectum of the midbrain.

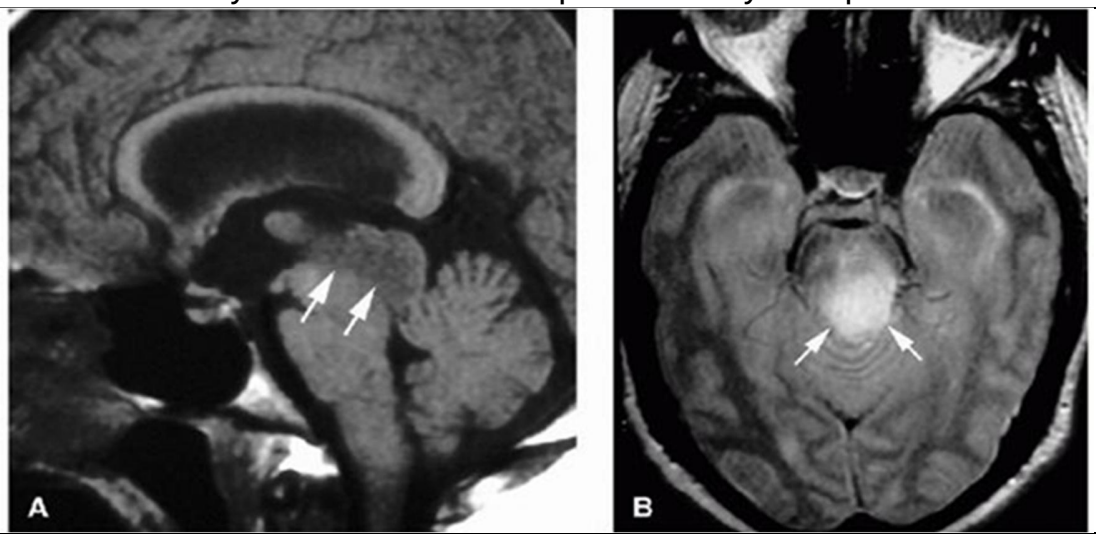
2 types:

- Well circumscribed (pilocytic, WHO grade I)
- Diffusely infiltrating low grade (WHO grades II).

- Age: Childhood

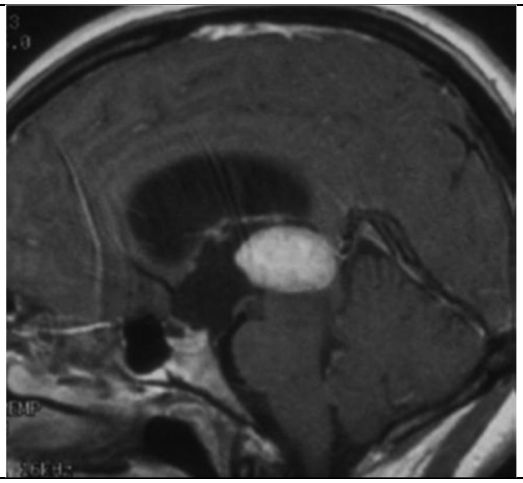
Imaging findings:

- Bulbous enlargement of the tectal plate is noted.
- T1: isointense on T1
- T2: hyperintense
- T1+C: no to minimal enhancement
- Secondary obstruction of the aqueduct => Hydrocephalus

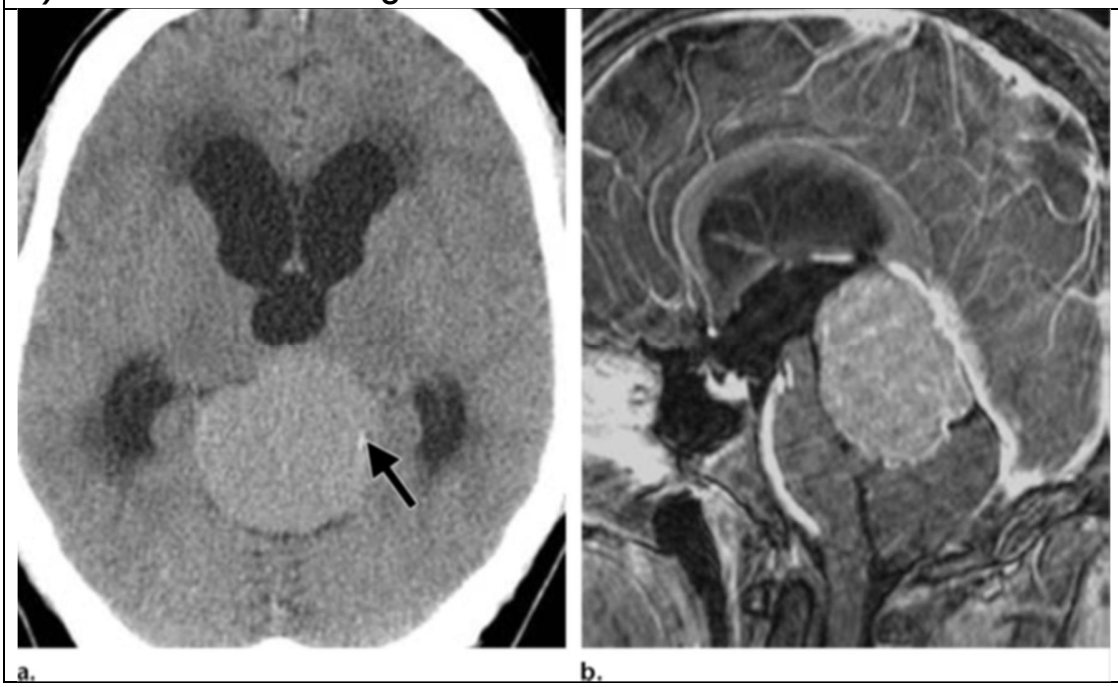


ii- Adult Pineal Lesions		
	Pineocytoma	Meningioma
WHO	Grade I	Grade I
Origin	Pineal gland	Tentorium
Age	Adult	
	Well defined lesion	Well defined lesion Dural based lesion
Ca	Exploding" pineal calcification toward periphery	Calcification: common
MRI	T1: Low T2: High	T1: Iso – low T2: Iso – High
+C	Heterogeneous enhancement	Homogenous enhancement Dural tail
Spread	No CSF dissemination	

1) Pineocytoma



2) Tentorial Meningioma



iii- Cystic Lesions

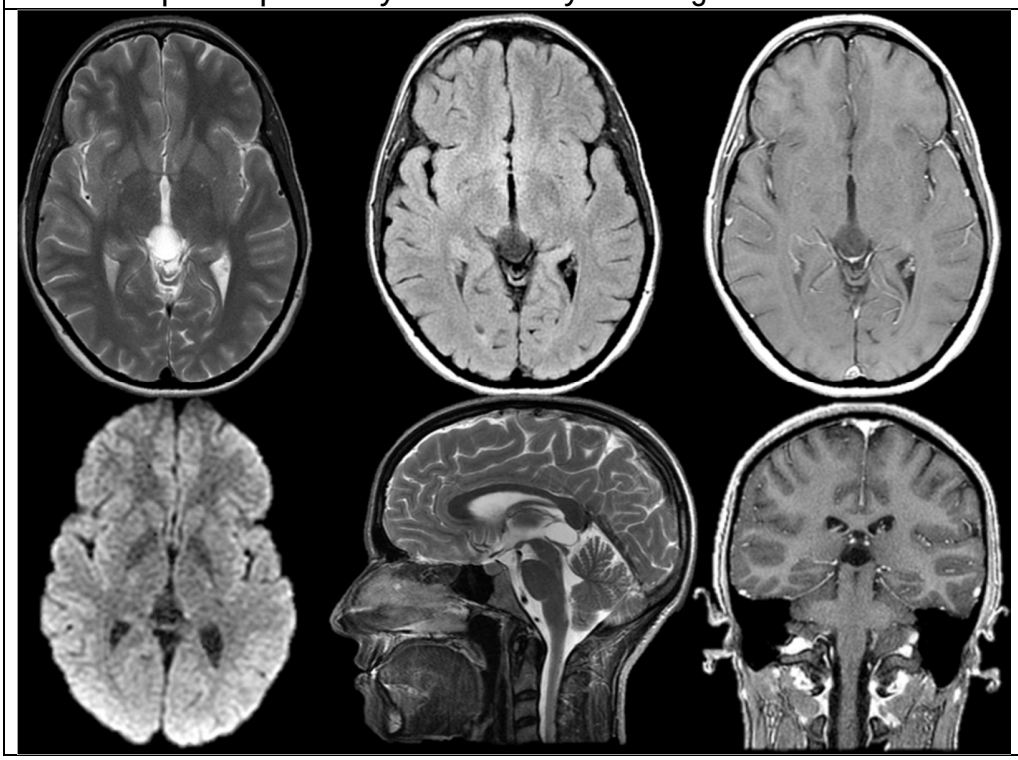
1) Pineal cyst
2) Arachnoid
3) Dermoid
4) Epidermoid
5) Quadrigeminal lipoma
6) Cavum septum interpositum
7) Aneurysmal dilation of vein of Galen

1) Pineal Cyst

- 25%–40% of cases in autopsy series
- 23% of patients in imaging studies of normal volunteers
- Age: occur in all age ranges
- typically asymptomatic
- 2–15 mm in size.
- When >>>15 mm, patients may become symptomatic, typically with headache or visual changes
- Intracystic hemorrhage (“pineal apoplexy”) and acute hydrocephalus rarely occur; resultant death has been reported

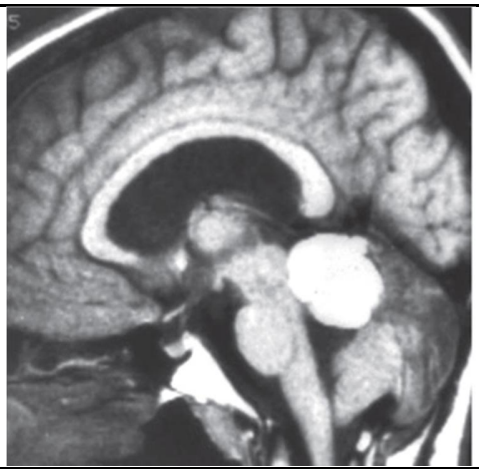
Imaging findings:

- Round or oval, thin-walled, and well-circumscribed.
- T1 & T2: CSF like signal
- FLAIR: the signal may not be completely suppressed due to the proteinaceous contents.
- T1+C enhancement of the cyst wall occurs in most pineal cysts but is typically incomplete; this finding has been attributed to fragmentation of the pineal parenchyma as the cyst enlarges



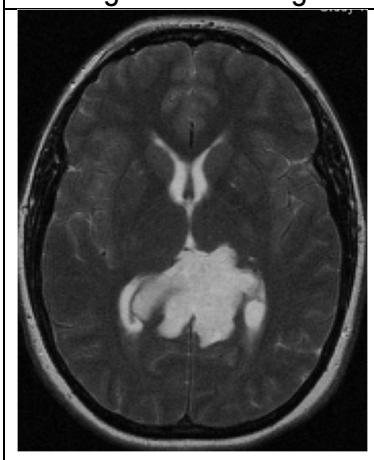
2) Quadrigeminal Lipoma

- CT: low attenuation, consistent with fat.
- MRI = fat
- T1:hyperintense
- T1 FS: saturation
- No enhancement

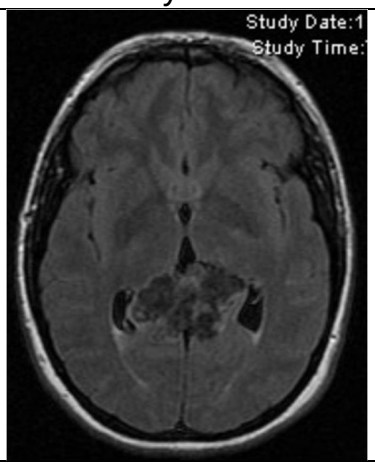


3) Quadrigeminal Epidermoid

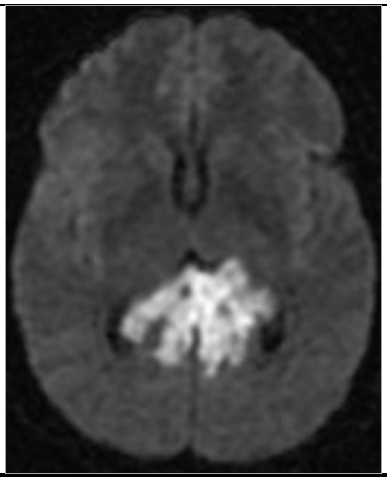
T2: High CSF like signal



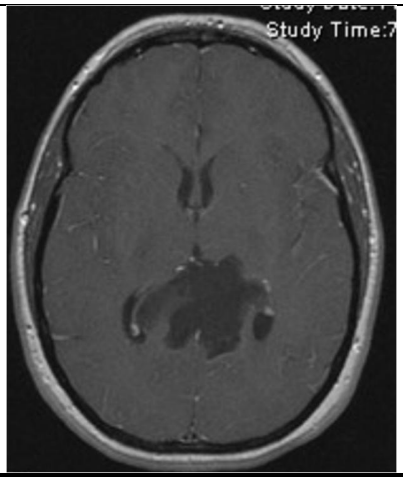
FLAIR: Dirty fat



DWI: Restricted



No enhancement



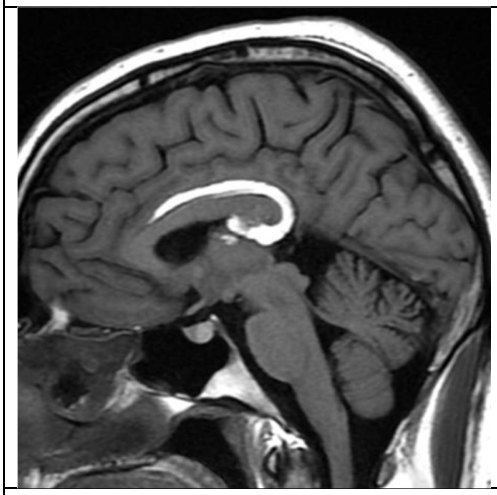
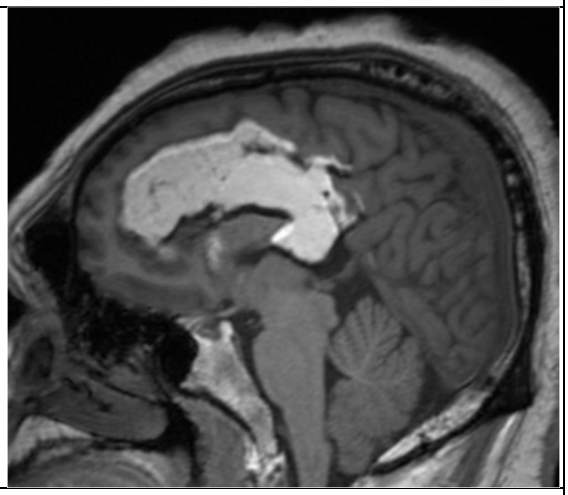
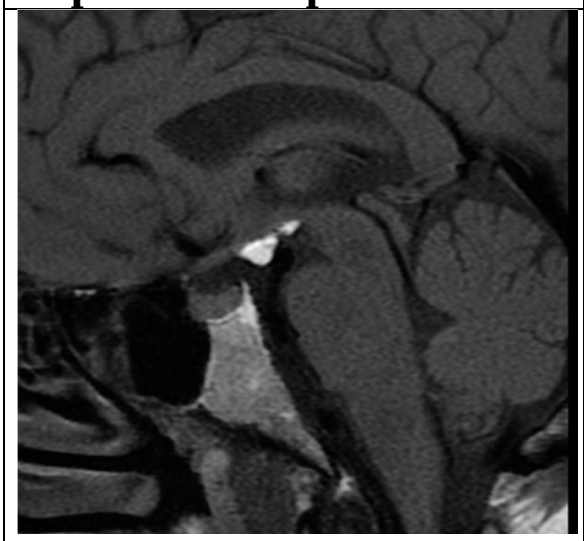
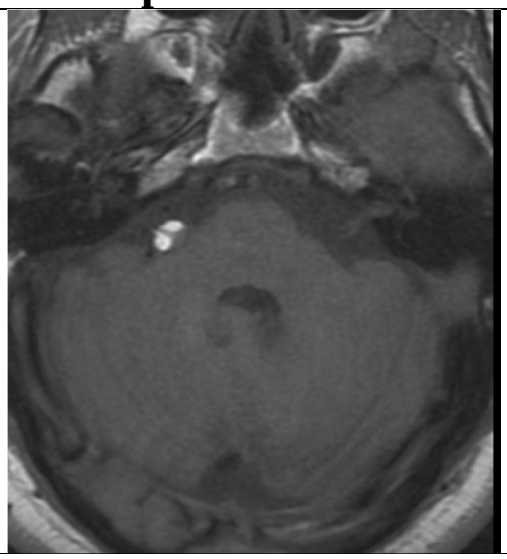
E- Extra-Axial Tumor Like lesions / Cystic Lesions Extra-axial Subarachnoid Lesions				
	Lipoma	Dermoid cyst	Epidermoid cyst	Arachnoid cyst
Site	Midline : 1) Pericallosal: Tubulonodular - Curvilinear 2) Quadrigeminal /Ambient cistern 3) Suprasellar cistern 4) CPA cistern 5) Spinal	Midline : 1) Parasellar 2) Frontobasal 3) Vermis 4) 4th Venticle 5) Spinal : Lumbosacral	Off-Midline : 1) CPA cistern 2) Parasellar 3) Middle cranial fossa 4) Diploic space	1) Middle cranial fossa 2) Suprasellar c. 3) Quadrigeminal 4) CPA 5)Cisterna magna 6) Cerebral convexity
CT	Fat density	- Fat density - Fluid density - Calcification	- CSF like density - lobulated borders	- CSF density - Smooth borders
MRI	*Fat signal* T1: High T2: High T1FS: Low	Fat signal Fluid signal Calcification	CSF like signal *FLAIR:* Intermediate (dirty) DWI: High	*CSF signal* T1: Low T2: High FLAIR: Low DWI: Low ADC: High
		Ruptured dermoid : show fat droplets in subarachnoid space or fat / CSF level in ventricles.	*Insinuate along CSF spaces*	
+C	*NO Enhancement*			

1) Intracranial Lipoma

Location

- Pericallosal lipoma (45%)
- Quadrigeminal cistern lipoma (25%)
- Suprasellar cistern lipoma (15%)
- Cerebellopontine angle lipoma (10%)
- Sylvian fissure (5%)

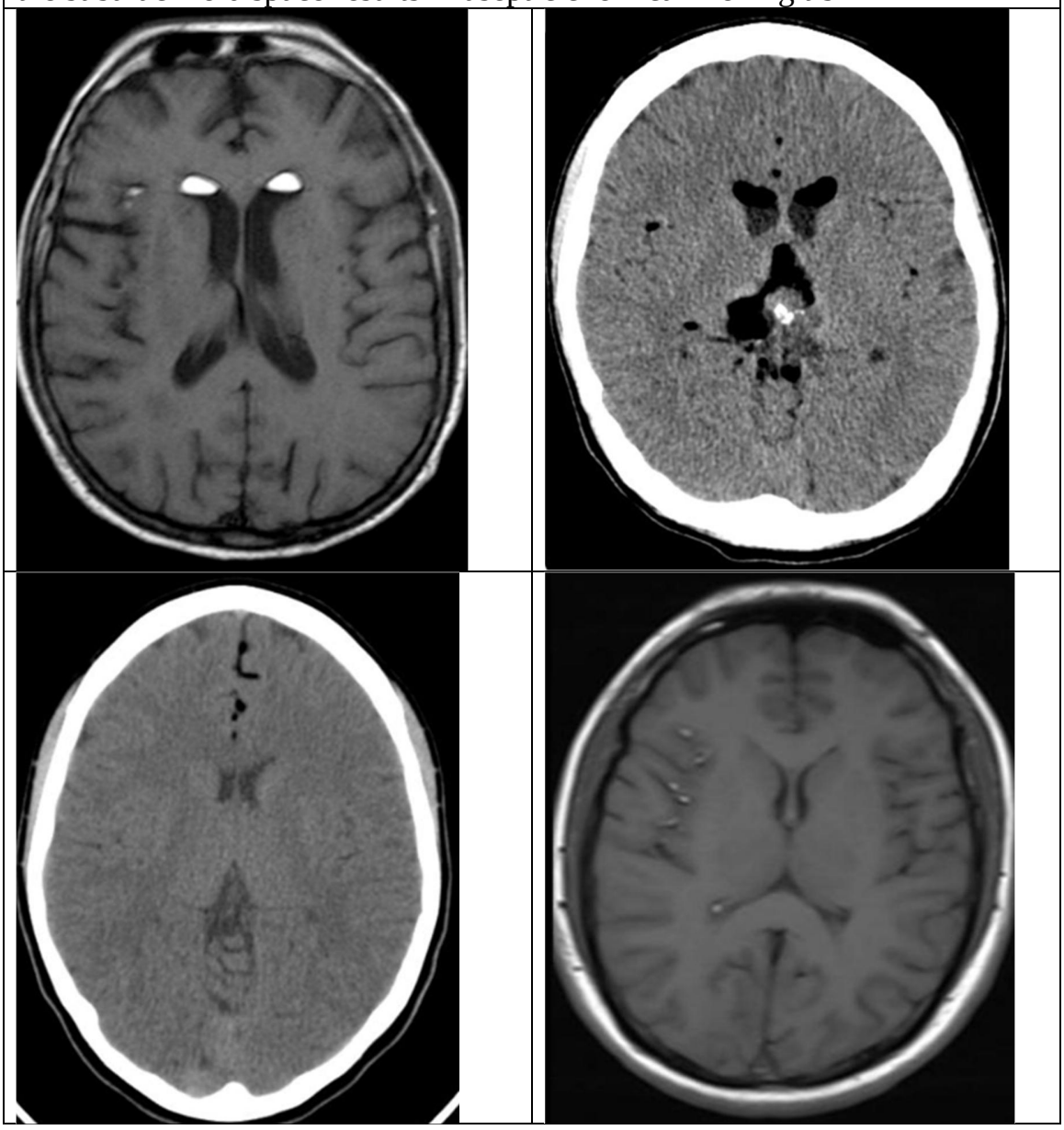
Pericallosal Lipoma ± Corpus Callosum Dysgenesis

Curvilinear	Tubulonodular
	
Suprasellar Lipoma	**CPA lipoma**
	

2) Intracranial Dermoid Cyst

❖ Rupture Dermoid

(spontaneous, traumatic, or iatrogenic (at resection)) leakage of sebum into the subarachnoid space results in aseptic chemical meningitis



3) Arachnoid Cyst

❖ Middle Cranial Fossa Arachnoid Cysts

❖ Galassi classification

Type I

- Small, spindle-shaped
- Limited to the anterior portion of the middle cranial fossa
- Below the spenoid ridge
- free communication of subarachnoid space

Type II

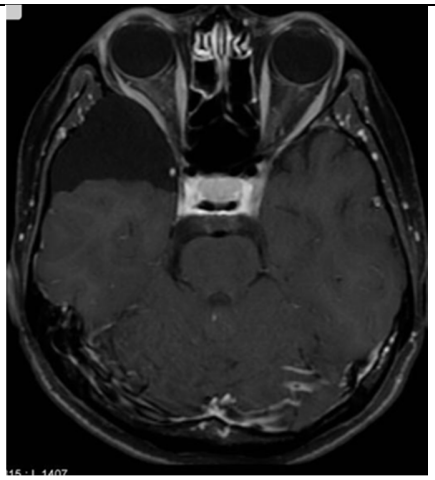
- Superior extent along ***sylvian fissure***
- Displacement of the temporal lobe
- Slow communication with subarachnoid space

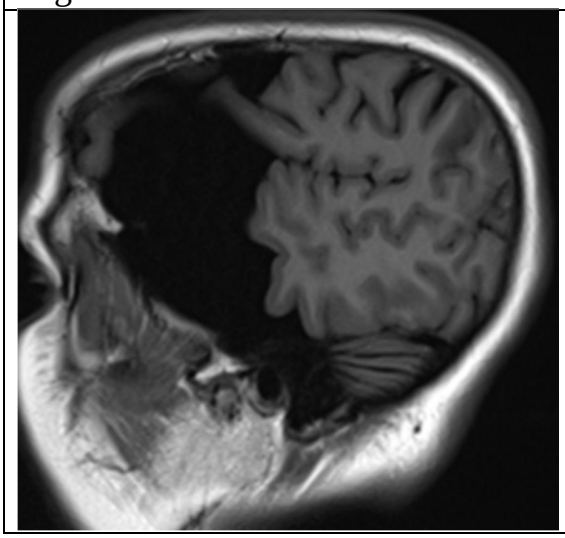
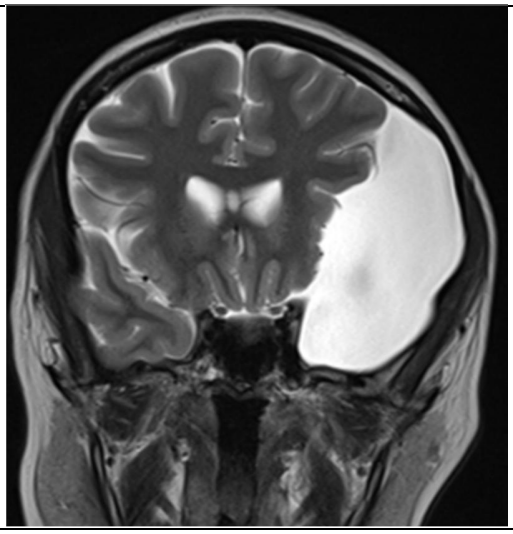
Type III

- Large, fills the whole middle cranial fossa
- Displacement of not only the temporal lobe but also the ***frontal and parietal lobes***
- often results in midline shift
- little communication with subarachnoid space

Type I Middle cranial fossa arachnoid cyst

Axial CT	Coronal CT	Sagittal CT

Type II Middle cranial fossa arachnoid cyst	
Axial T1: Low signal	Axial T2: high signal
Axial FLAIR: Low signal	Axial T1+C FS: No enhancement
	
Axial DWI: facilitated	Axial ADC: High signal

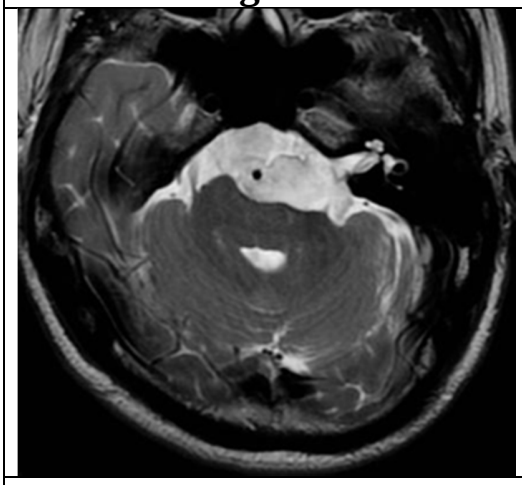
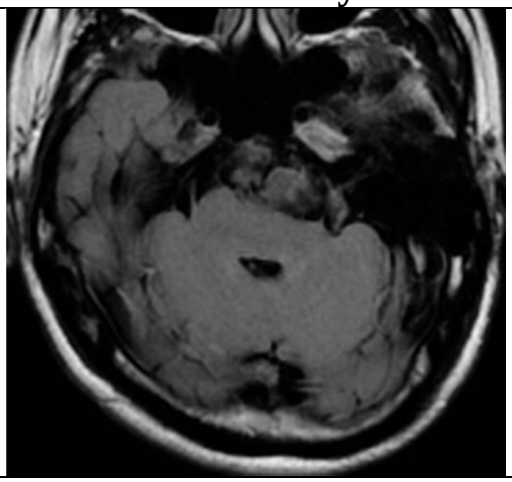
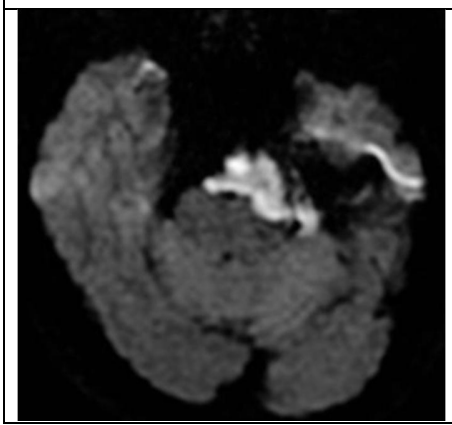
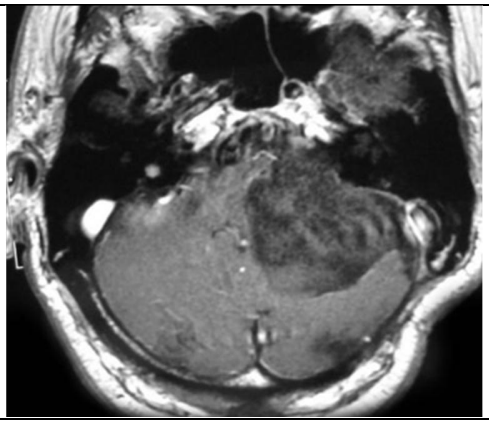
Type III Middle cranial fossa arachnoid cyst	
Sagittal T1	Coronal T2
	

❖Posterior Fossa Arachnoid Cyst	
Retrocerebellar	**CPA**

4) Intracranial Epidermoid

Location

- Intradural: 90%
  - Cerebellopontine angle: 40-50%
  - Suprasellar cistern: 10-15%
  - Fourth ventricle: ~17% 8
  - Middle cranial fossa
  - Interhemispheric
  - Spinal (rare)
- Extradural: 10%
  - Most within skull

Axial T2: High	Axial FLAIR: Dirty
	
Axial DWI: Restricted	**+C: No enhancement**
	

F- Diffuse Meningeal Lesions

1) Infection :
   a) Viral – Bacterial – TB – Fungal meningitis
   b) Subdural / Epidural abscess - empyema
2) Inflammation :
   a) Tuberculosis
   b) Neurosarcoidosis
   c) Wegner granulomatosis
   d) Langerhans cell histocytosis
3) 1ry Neoplastic :
   a) Meningioma – Hemangiopericytoma – Sarcoma – Melanoma
4) Meningeal carcinomatosis :
   a) CNS tumors :
      i) Posterior fossa: Medulloblastoma – Ependymoma
      ii) Pineal region: Germinoma – Pineoblastoma
   b) Non CNS tumors :
      i) 2ry CNS Lymphoma – Leukemia
      ii) Breast – Prostate cancer
5) Trauma / Vascular :
   a) Chronic subdural hematoma
   b) Subarachnoid hemorrhage

1) Meningioma

- Most common extra-axial tumours of the central nervous system.
- Non-glial neoplasm originates from the arachnoid cap cells of meninges

- Age : 40 – 60 y Sex : F > M
- Multiple: 5-10%; Associations: NF-II

WHO Grade:

- ❖ WHO Grade I = Typical meningioma (88-95%)
- ❖ WHO Grade II = Atypical meningioma (5-6%)
- ❖ WHO Grade III = Malignant meningioma ~1%
- ❖ WHO Grade IV = Meningioma with sarcomatous degeneration (extremely rare)

Location:

- o 85-90% supratentorial
  - 45% Parasagittal, convexities
  - 15-20% Suprasellar
  - 10% Olfactory groove
  - 5-10% Parasellar
- o 5-10% infratentorial
- o <5% miscellaneous intracranial
  - Intraventricular meningioma (choroid plexus)
  - optic nerve meningioma
  - Pineal gland
  - Spinal: especially thoracic (see spinal meningioma)

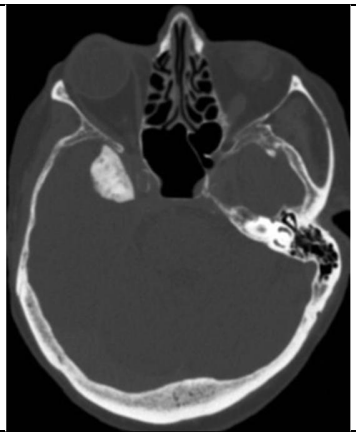
Imaging Findings:

- Well defined extra-axial meningeal based lesion
- NCCT: hyperdense ± Calcification (25%)– Bone hyperostosis
- T1: Iso – Low
- T2: Iso (50%) – Hyper (40%) – Hypo (10%)
- T1+C: Intense homogenous enhancement + Dural tail

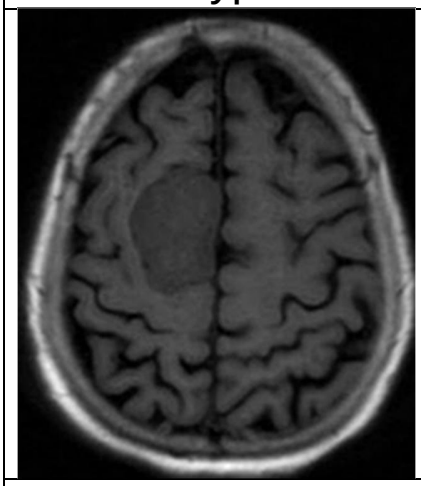
Angiography :

- Spokewheel appearance
- Dense venous filling
- Mother in law = Comes early + Stays late
- AV shunting + Early draining veins

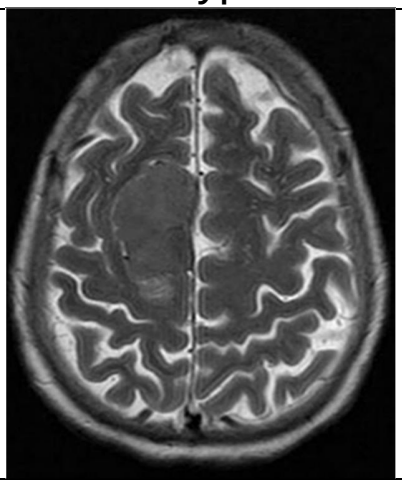
NCCT: Extra-axial + Hyperdense + Calcification



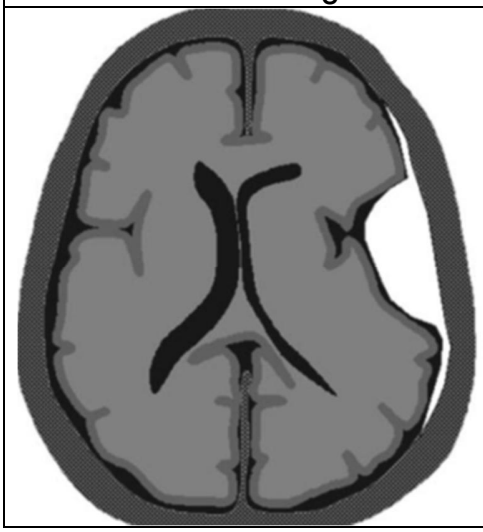
T1: Iso - Hypo

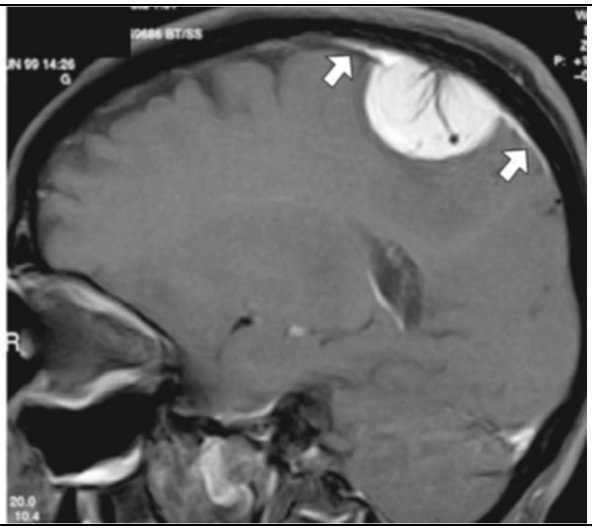


T2: Iso - Hypo



+C: Intense homogenous enhancement + Dural tail





❖DWI & ADC

⇨ ADC cut off value = 0.8X10-3mm/sec

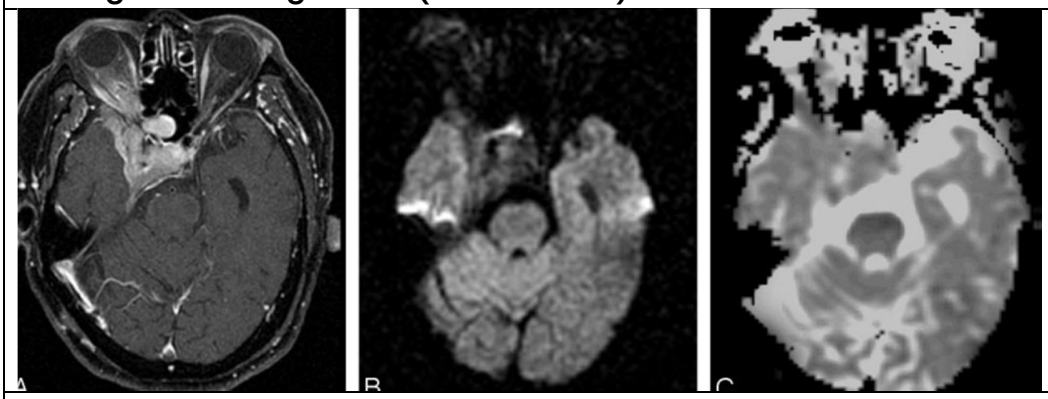
Benign Meningioma (WHO Grade I)

- DWI = Isointense
- ADC map = Isointense
- ADC = 0.8-1.8X10-3mm/sec

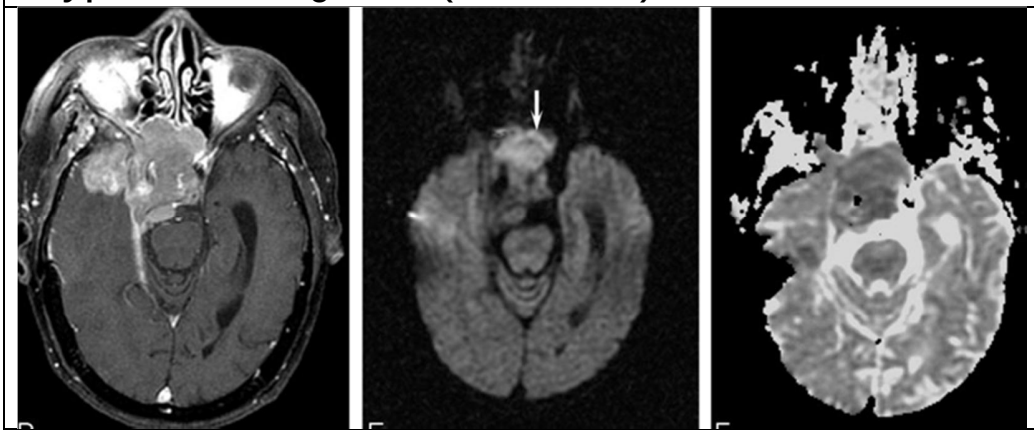
Atypical / Malignant Meningioma (WHO Grade II /III)

- DWI: Hyperintnese
- ADC map = Hypointense
- ADC= 0.45-0.69 X10-3mm/sec

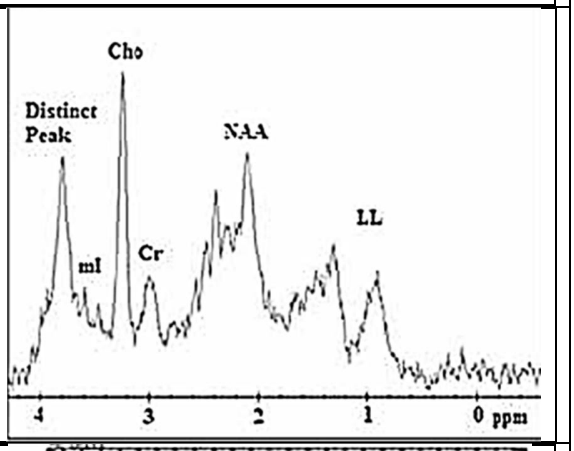
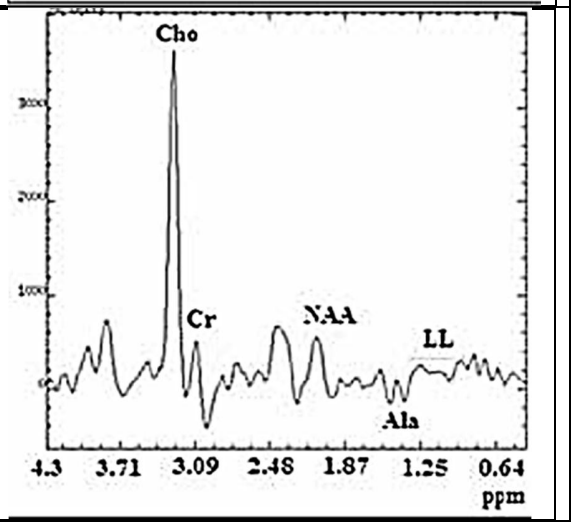
Benign meningioma (ADC = 0.9)



Atypical meningioma (ADC = 0.6)



❖ MRS

Short TE	Long TE
• Increased Alanine (Doublet or Triple peaks at 1.3-1.5 ppm) at short TE with inversion at long TE • Distinct peak (?Glx) at 3.8ppm => discriminating meningiomas from high-grade gliomas and metastases. • Increased glutamate-glutamine complex (Glx) multiplet signal at 2.35ppm	• Increased Choline (Cho): cellular tumor. • NO or low N-acetylaspartate (NAA): non-neuronal origin • NO or low creatine (Cr) • Highest Cho/Cr ratio • Highest Cho/NAA ratio
<u>Meningioma at short TE</u> o Distinct peak at 3.8ppm o Alanine doublet peak at 1.3ppm o Glx multiplet signal at 2.35ppm	
<u>Meningioma at long TE</u> o Elevated Cho at 3.2ppm o Absence of NAA at 2.02 ppm o Ala doublet inversion at 1.47 ppm	

2) Meningeal Haemangiopericytoma

- They were previously classified as angioblastic sub-type meningiomas.
- These tumours arise from smooth muscle perivascular pericytes of dural capillaries (pericytes of Zimmerman).
- They are more aggressive than meningiomas
- Higher frequency of recurrence
- WHO grade II tumour

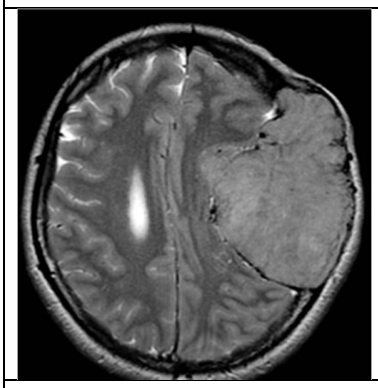
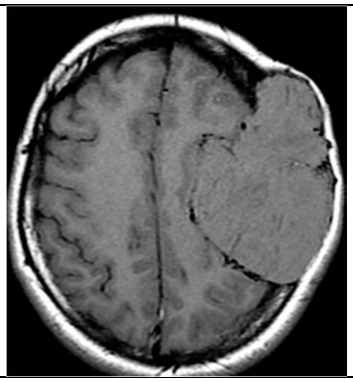
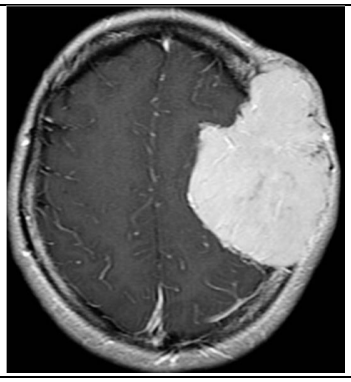
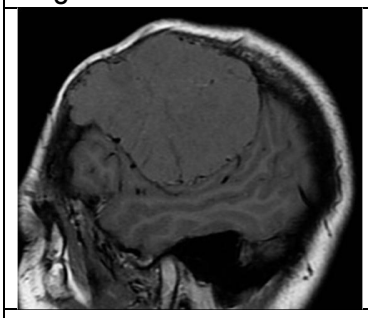
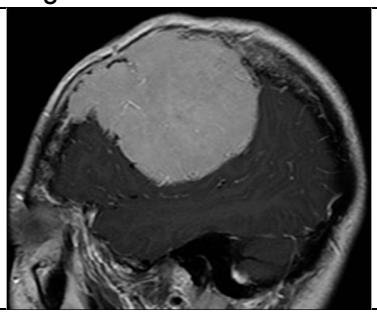
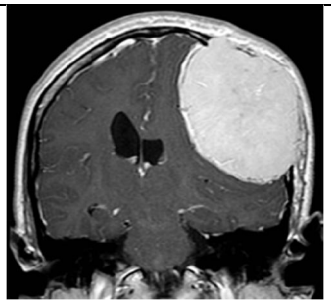
Imaging findings:

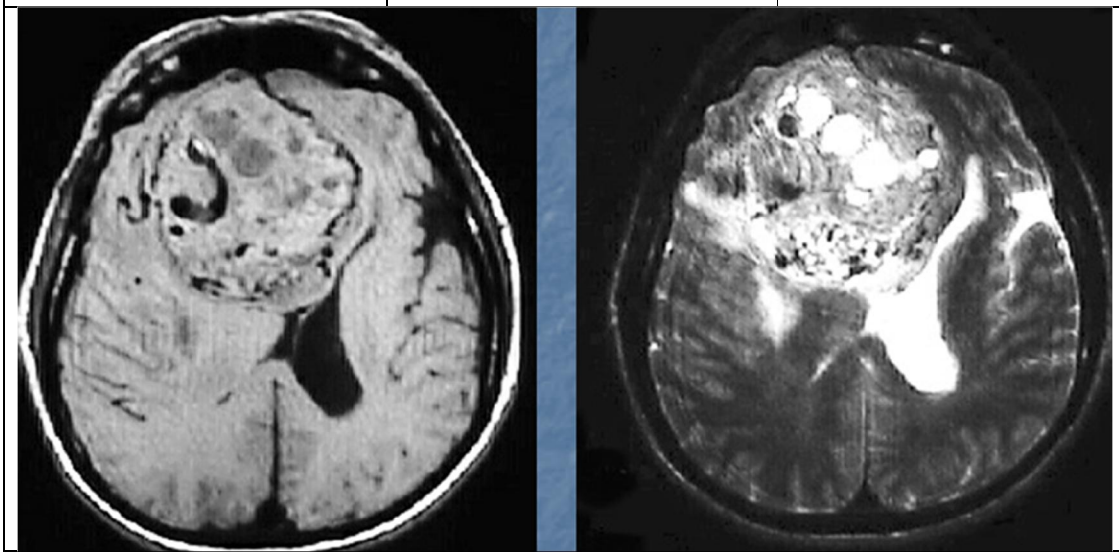
- Distinguishing a haemangiopericytoma from a meningioma can be difficult as they have similar appearances on both CT and MRI.

Helpful features include :

- lobulated contour
- absence of calcification and hyperostosis
- invasion of the skull (common)
- multiple flow voids on MRI
- Corkscrew arteries
- may have a narrow base of dural attachment

	HPC	Meningioma
ADC	1.116 ± 0.127	0.875 ± 0.014
MRS	• High myo-inositol peak at 3.56 ppm • Lack of alanine	• Low myo-inositol peak at 3.56 ppm • Alanine peak

Axial T2	Axial T1	Axial T1+C
		
Sagittal T1	**Sagittal T1+C**	**Coronal T1+C**
		



2) Meningeal Carcinomatosis

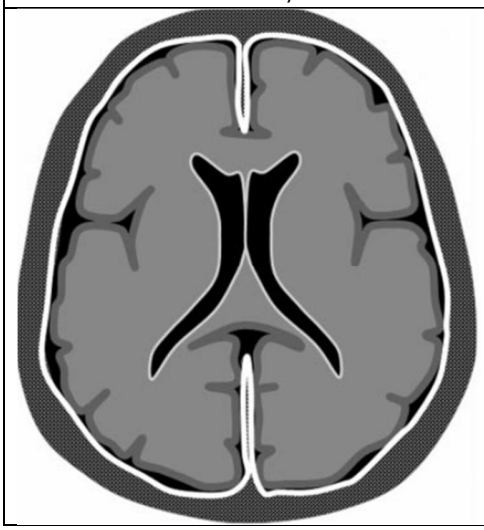
Origin of meningeal carcinomatosis:

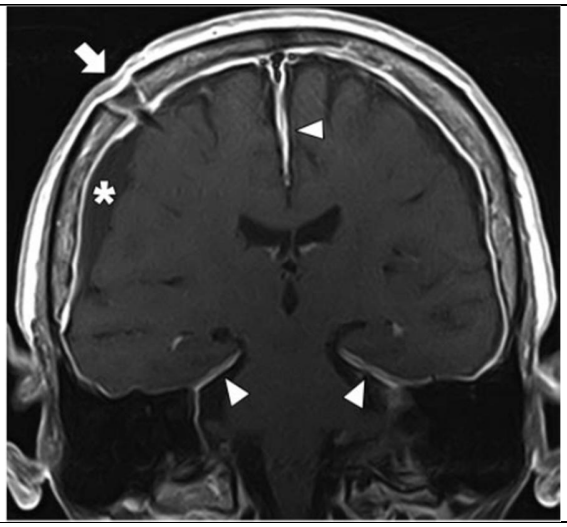
- CNS tumors :
  - Posterior fossa: Medulloblastoma – Ependymoma
  - Pineal region: Germinoma – Pineoblastoma
- Non CNS tumors :
  - 2ry CNS Lymphoma – Leukemia
  - Breast – Prostate cancer
- DD: Infection :
  - Viral – Bacterial – TB – Fungal meningitis
- DD: Inflammation :
  - Tuberculosis
  - Neurosarcoidosis
  - Wegner granulomatosis
  - Langerhans cell histocytosis

2 patterns:

a. Pachymeningeal (thick meninges) enhancement:

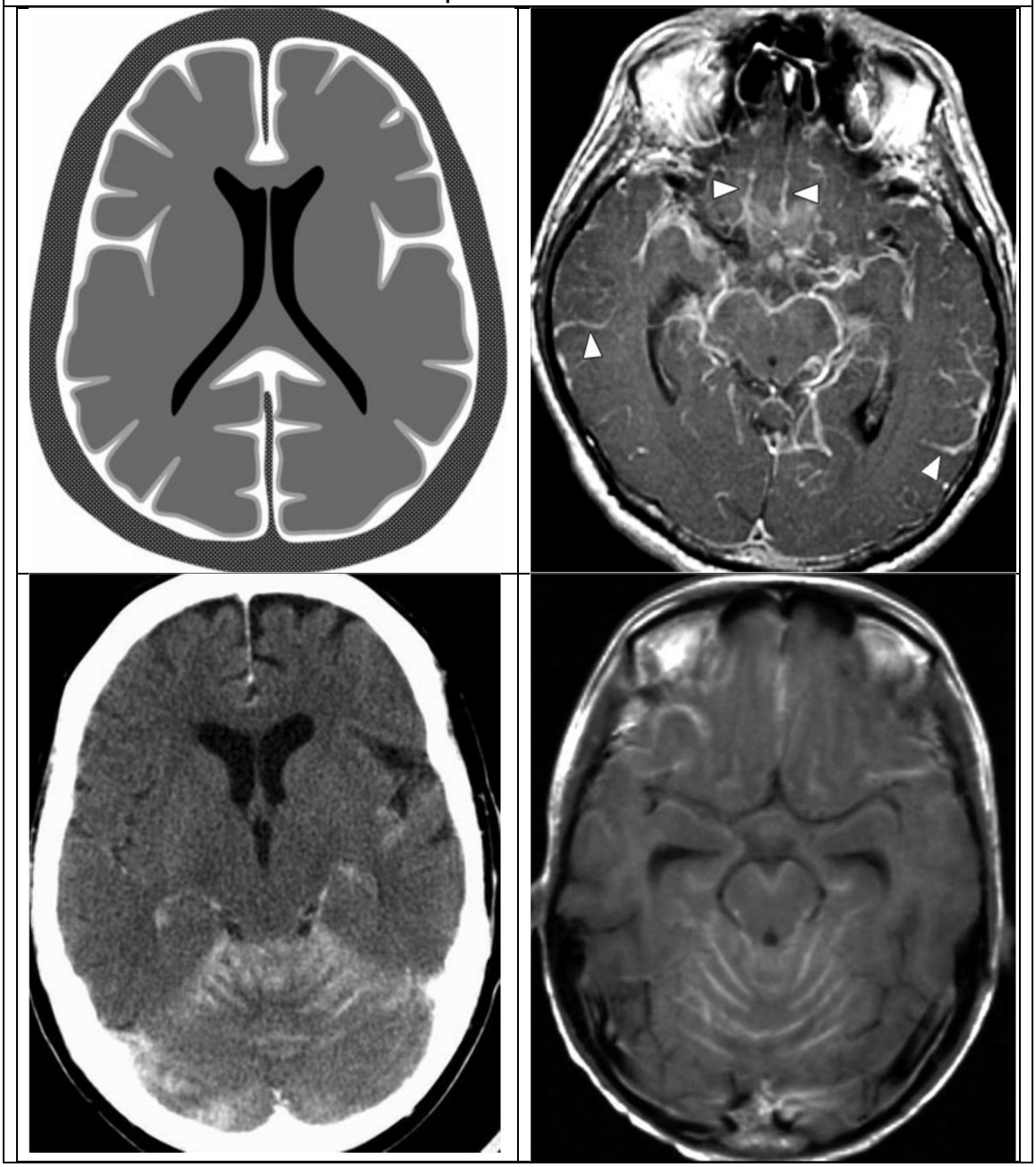
- Abnormal dural enhancement: Thick, nodular, continuous
- Normal dura enhancement: Thin, linear, and discontinuous
- Along the inner table of the shull
- Dural reflections of the falx cerebri, tentorium cerebelli, falx cerebelli, and cavernous sinus





b. Leptomeningeal (skinny meninges) enhancement

- Pia-arachnoid
- Along the surface of the brain
- "gyriform" or "serpentine" appearance.
- fills the subarachnoid spaces of the sulci and cisterns



G- General Calvarial and Skull Base Bone Lesions

<table>
<tr><th colspan="2">1ry</th><th>2ry 3M + 3L</th></tr>
<tr><th>Benign</th><th>Malignant</th><td rowspan="5">Metastasis
Multiple myeloma / Plasmacytoma
Melanoma

Leukemia
Lymphoma
LCH / EG</td></tr>
<tr><td rowspan="4"><u>Sclerotic Lesions:</u>
Osteoma
Osteoblastoma
Ossifying fibroma
Fibrous dysplasia
Intraosseous meningioma
Paget disease
Hyperostosis Frontalis Interna
Osteopetrosis
Haemangioma
Thalassemia</td><td>Chordoma
chondrosarcoma</td></tr>
<tr><td>Osteosarcoma
Ewing sarcoma</td></tr>
<tr><td rowspan="2"></td></tr>
<tr><td>Giant cell tumour (GCT)
Aneurysmal bone cyst (ABC)</td><td></td></tr>
<tr><td>Epidermoid
Dermoid cysts</td><td></td></tr>
</table>

1) Fibrous Dysplasia

- Fibrous dysplasia (FD) is a benign tumour-like congenital process, manifested as a defect in osteoblastic differentiation and maturation, with progressive replacement of normal bone with immature woven bone

- Age: Young affected (< 30 years old)
- F>M = 3:1
- Location: Affects any bone including skull, skull base & facial bones
- Bone CT: increased bone volume with "ground glass" appearance
- MRI: Expansile lesion with low signal on both T1 & T2 images

4 Types:

- Monoostotic : single bone
- Polyostotic : multiple bones
- Craniofacial fibrous dysplasia : skull and facial bones alone
- Cherubism : mandible and maxilla alone (not true fibrous dysplasia)

2 Associations

- Mazabraud Syndrome
- McCune-Albright Syndrome

3 Presentations:

- Monostotic FD
  - 70% of all FD cases; single osseous site is affected
  - Older children & young adults (75% present before age 30)
  - Skull base & face involved in 25%; maxilla (especially zygomatic process) & mandible (molar area) > > frontal bone> ethmoid & sphenoid bones> T-bone
  - May be asymptomatic, incidental imaging findings
  - Other symptoms nonspecific: Pain, focal swelling & tenderness
- Polyostotic FD
  - 25% of all FD cases; involves ≥ 2 separate osseous sites
  - Skull base & face involved in 50%
  - Younger patient group, mean age at diagnosis of 8 years
  - 2/3 have symptoms by age 10 including craniofacial asymmetry
- McCune-Albright syndrome
  - 5% of FD cases
  - Clinical triad of polyostotic FD (usually unilateral), endocrine

dysfunction (with precocious puberty) & cutaneous hyperpigmentation (cafe-au-lait spots)
  - Appears earlier & affects more bones more severely

3 CT features:

- Pagetoid (mixed) pattern (50%): Mixed radiopacity & radiolucency
- Sclerotic FD (25%): "Ground-glass" density
- Cystic FD (25%): Centrally lucent lesions with thinned but sclerotic borders

MRI features:

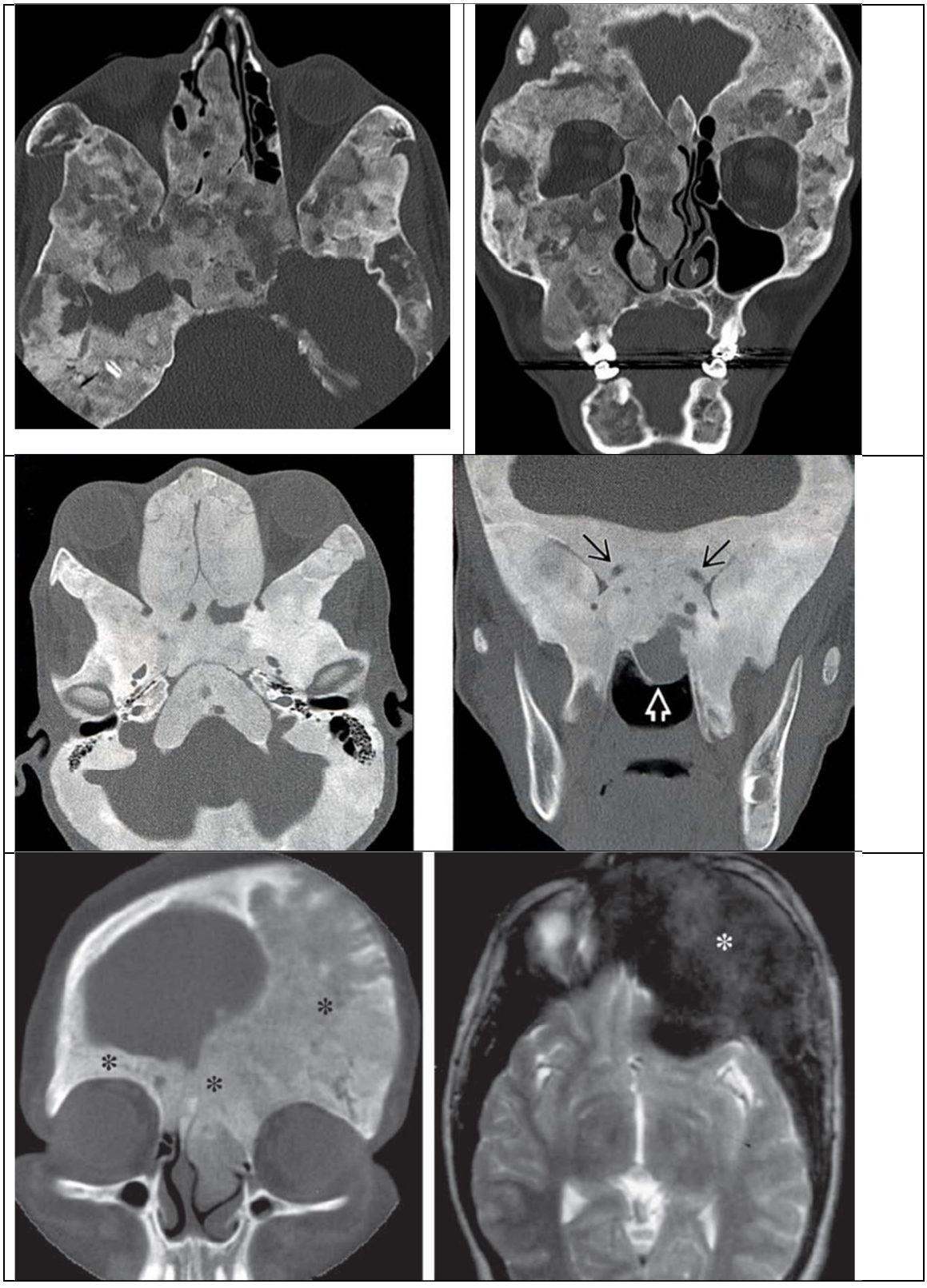
T1 : heterogeneous iso - Low
T2 : heterogeneous iso - low
T1 C+ (Gd) : heterogeneous contrast enhancement

Plain Xray:

- Well defined medullary lesion
- Medullary expansion and widening of the diploic space (both inner and outer table are affected)
- Endosteal scalloping
- Intact cortex
- No periosteal reaction
- Bone Deformity:
  - Bowing deformity
  - Shepherd's crook deformity of femoral neck
  - Discrepant limb length
  - Protrusio acetabuli

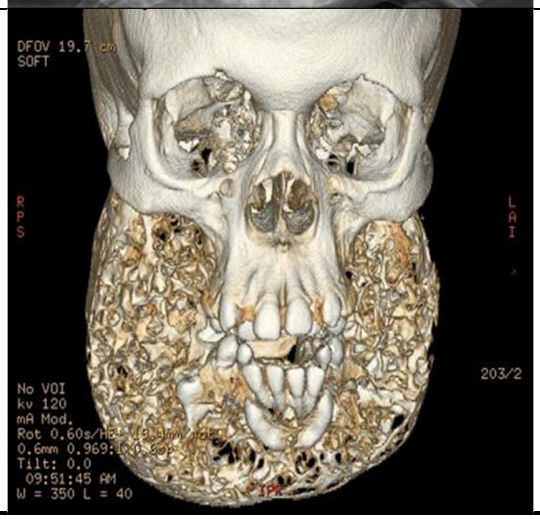
❖ Craniofacial FD = Leontiasis ossea:

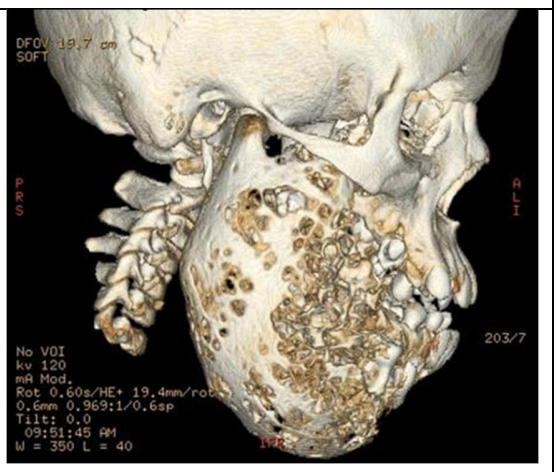
- is one of 4 types of fibrous dysplasia
- is characterized by involvement of the skull and facial bones.
- The craniofacial bones are affected in up to 50% of polyostotic cases.
- Occasionally it is seen in the setting of McCune-Albright syndrome



❖ Cherubism

- Bilateral expansile multiloculated cystic masses with symmetric involvement of mandible and maxilla
- Radiographic features consists of lucent expanded regions within the maxilla and mandible, with soap-bubble appearance.





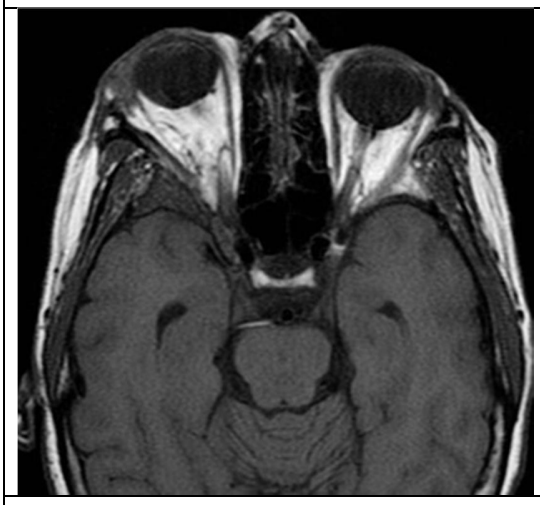
2) Intraosseous Meningioma

- is a sub type of meningioma.
- It is a rare tumour and account for less than 1% of all osseous tumours.

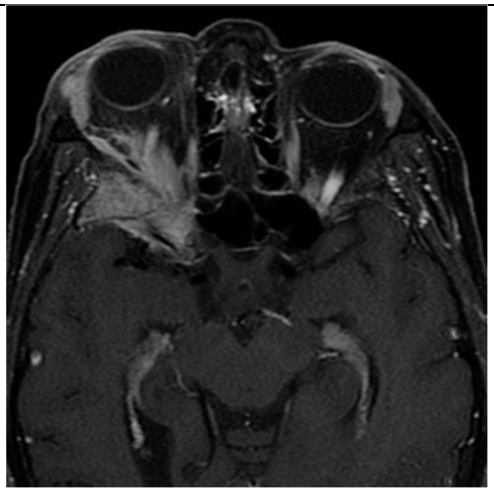
Imaging findings:

- Focal sclerotic bony lesion
- Enhancing extra-osseous soft tissue component
- Dural tail enhancement

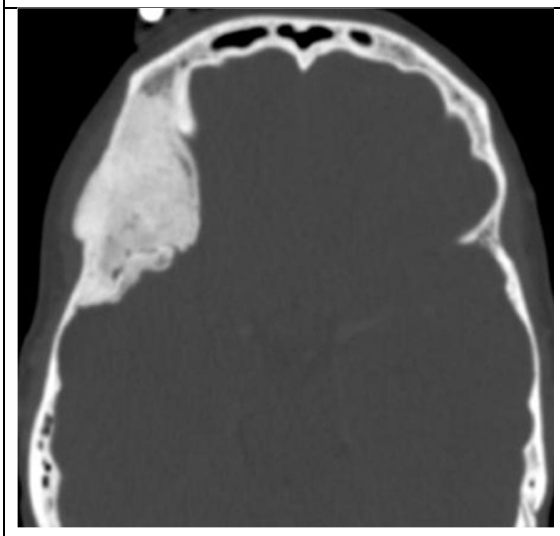
Axial T1



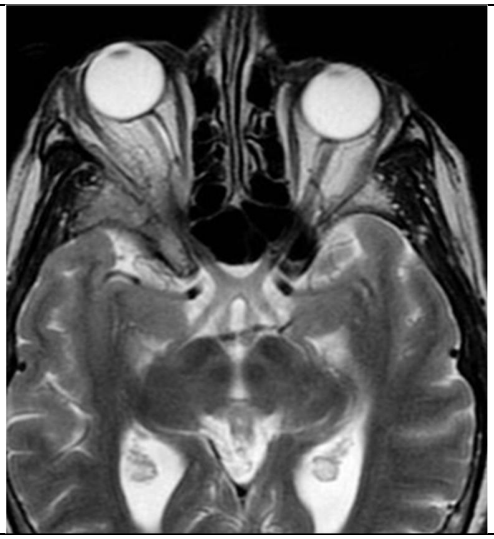
Axial T1 +C



Axial CT



Axial T2



3) PAGET DISEASE

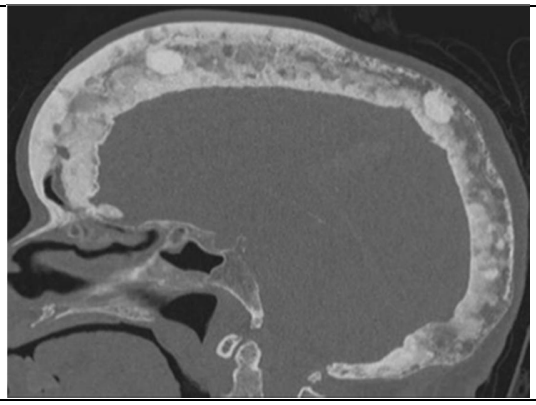
- Bone dysplasia characterized by excessive remodeling of bone resulting from alternating waves of osteoclastic & osteoblastic activity.

- Age > 40y
- M > F = 4: 1

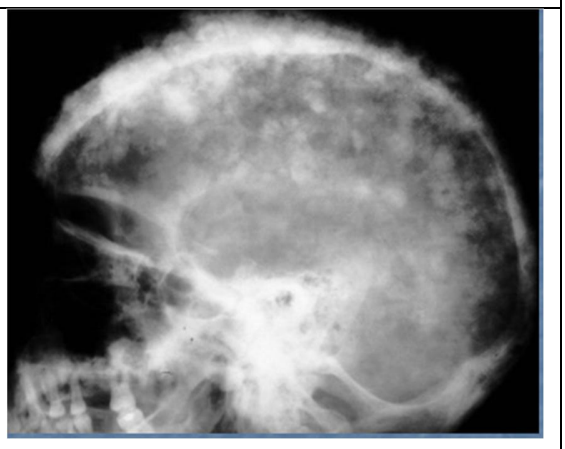
- Calvarium & cranial base: Diffuse thickening with "cotton-wool" appearance

- Early phase: Demineralization
- Intermediate phase: "Ground glass" appearance
- Late phase: Extreme bone thickening with diploic heterogeneity

Tam O'Shanter sign = Mosaic Pattern

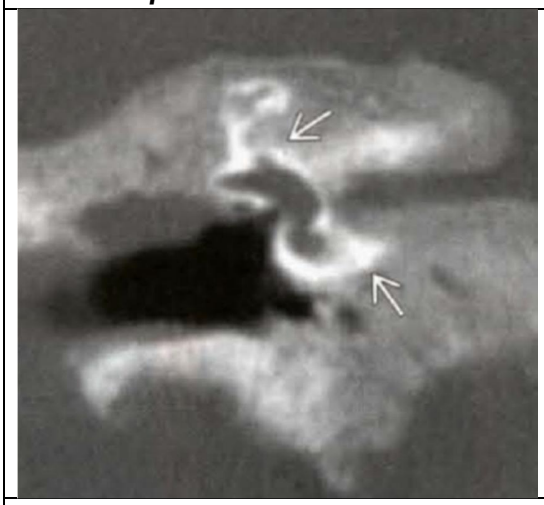


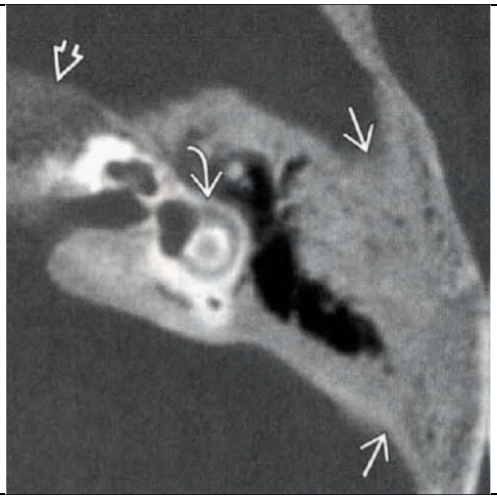
Cotton Wool Appearance



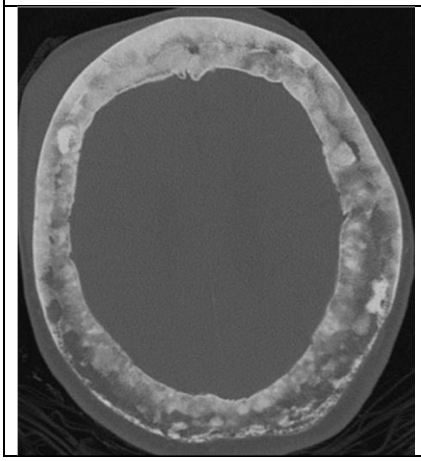
Paget Osteoporosis Circumscripta Cranii

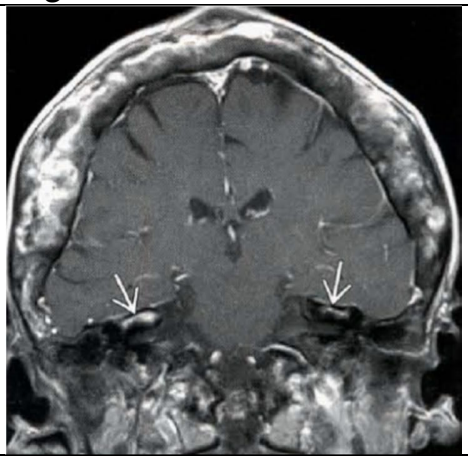
Petrous bone Ground Glass appearance +
Otic capsule demineralization





Diffuse calvarial thickening + heterogeneous enhancement



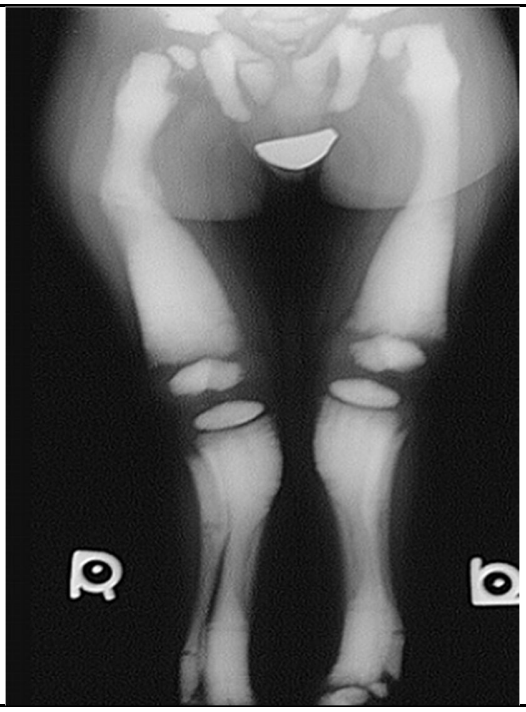


4) OSTEOPETROSIS

- Rare heritable metabolic bone disease with defective bone remodeling
- Autosomal Recessive Osteopetrosis (AROP): Childhood form; severe osteopetrosis
- Autosomal Dominant Osteopetrosis (ADOP): Adult form; less severe osteopetrosis

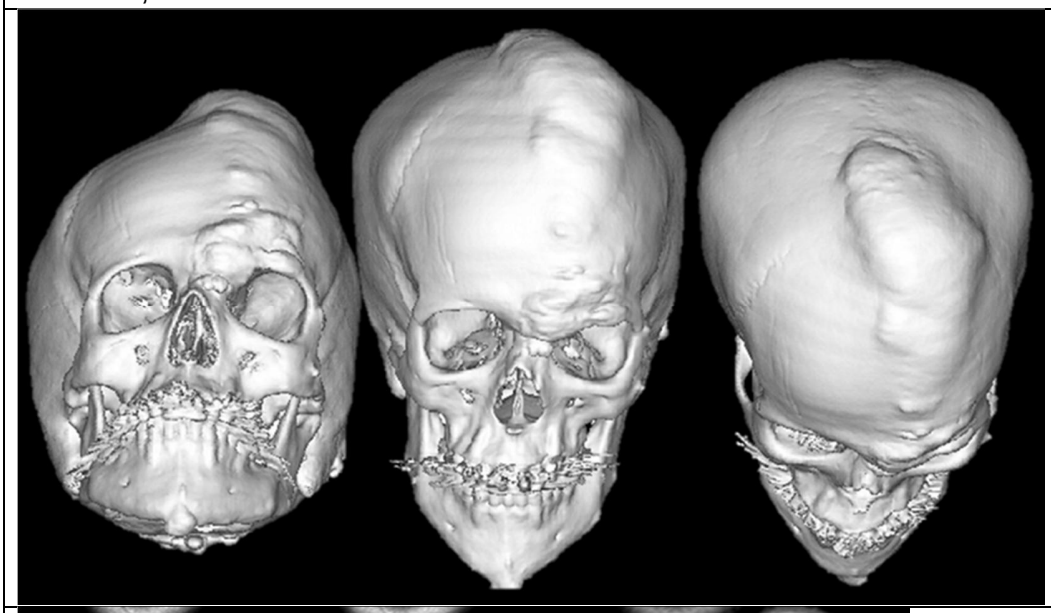
<u>Imaging findings:</u>

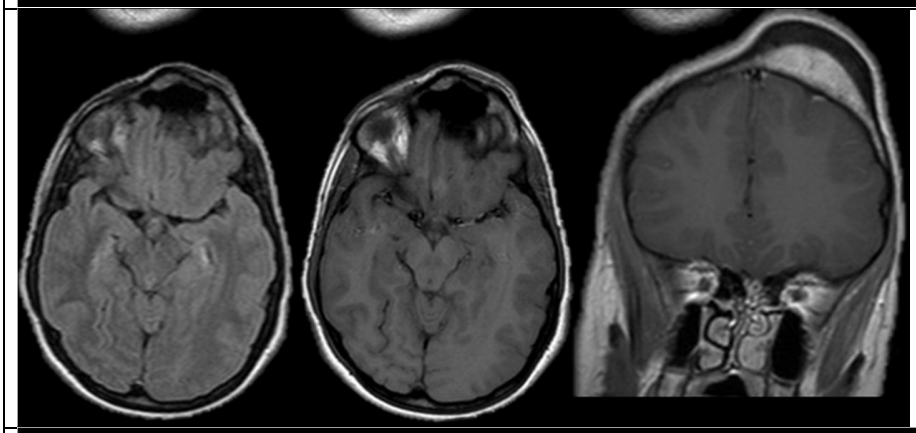
- Macrocranium
- Bone within bone appearance
- Cortical thickening with medullary encroachment
- Thick dense skull base + calvaria
- Obliteration of mastoid cells, paranasal sinuses,
- Narrowed basal foramina leading to CR nerve palsy
- Proptosis, hypertolerism

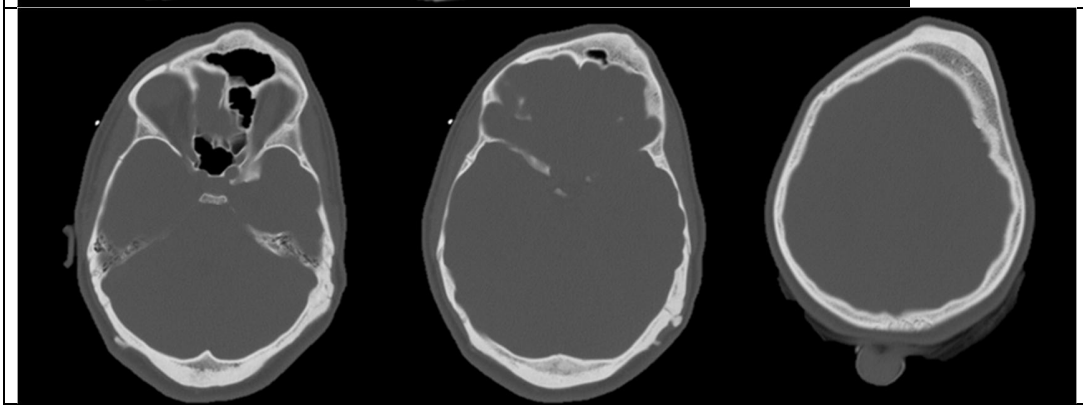


5) Proteus Syndrome = Elephant man Syndrome

- Rare genetic disorder causing hamartomatous overgrowth of the skin, bone, muscle and vessels







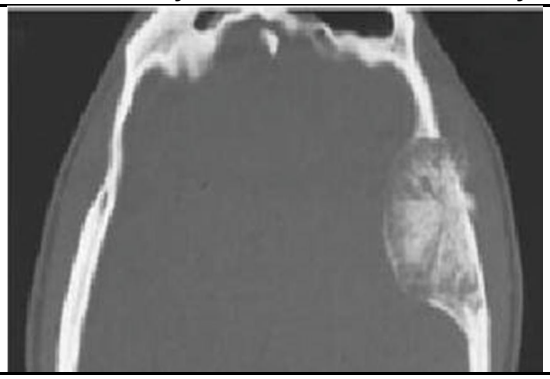
6) Intraosseous Hemangioma

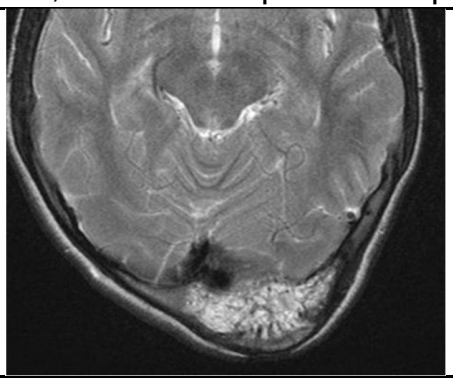
<u>Location specific sub types</u>

- Vertebral haemangioma
- Sacral haemangioma
- Skull vault haemangioma
- Intracortical haemangioma

<u>Imaging findings:</u>

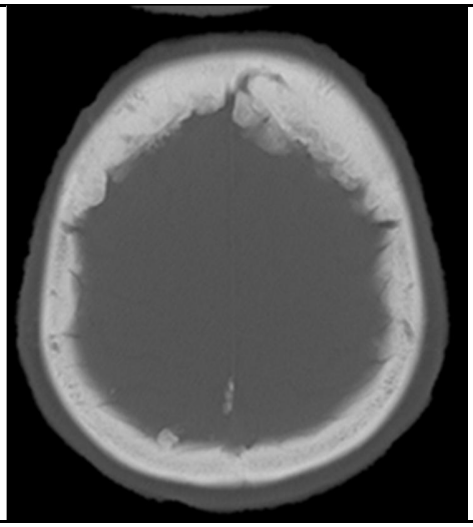
Internal bony trabeculation in honeycomb, sunburst or spokewheel pattern





7) Hyperostosis Frontalis Interna

- Benign bony overgrowth of the inner table of the frontal bone
- Incidental asymptomatic findings
- Most common in elderly females



7) LANGERHANS Cell HISTOCYTOSIS (LCH)

A. Eosinophilic Granuloma (EG)

- Mildest form of LCH
- Age of onset: Children> 5 years old
- *Unifocal*, osseous lesions only
- Long bones, skull, skull base & temporal bone
- Prognosis: Excellent

B. Hand-Schuller-Christian disease

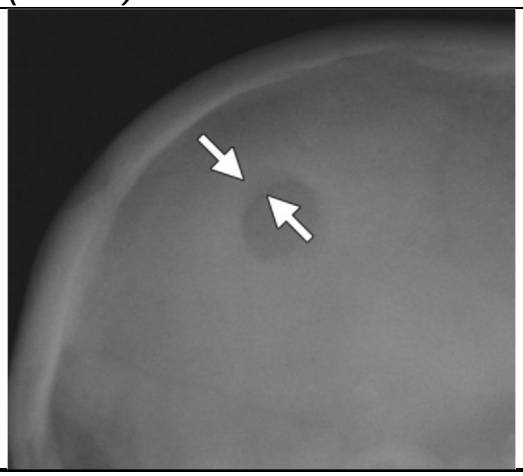
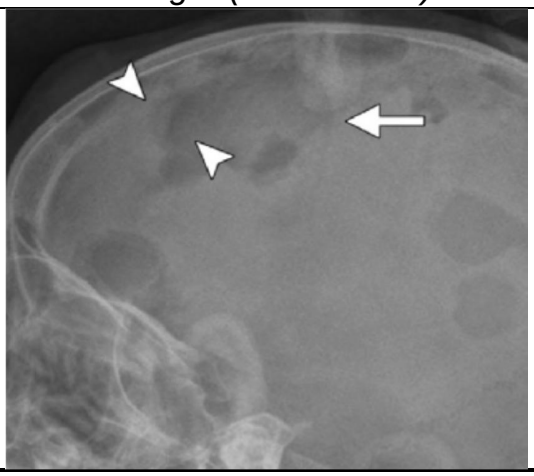
- More severe form LCH
- Age of onset: Children 1-5 years old
- *Multifocal* bone, skin, viscera and brain
- Classic triad of diabetes insipidus, proptosis & skeletal involvement
- Prognosis: Good

C. Letterer-Siwe disease

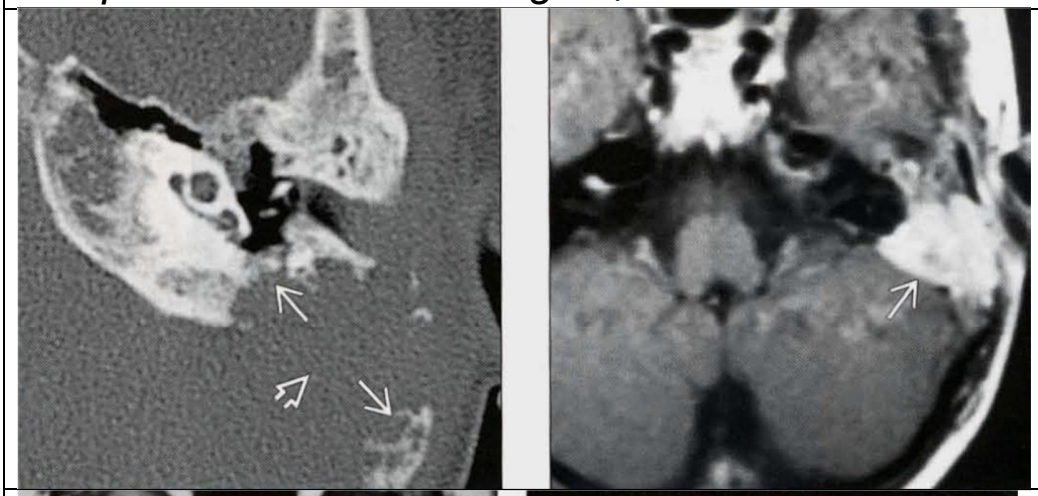
- Most severe form of LCH
- Age of onset: 0-3 years old
- *Fulminant disseminated* multi-organ disease including liver, spleen, lymphatic, lung & bone
- Prognosis: Poor with high mortality rate

Imaging findings:

- Well defined Lytic *"punchedout" skull lesion* with characteristic beveled edge; caused by asymmetric destruction of the inner and outer cortices
- Skull lesions may grow in size and coalesce, creating a maplike appearance referred to as a *geographic skull*

Lytic "punchedout" skull lesion with a characteristic beveled edge (arrows).	*large coalesced lesions (arrow), "geographic skull," with classic beveled edges (arrowheads).*
	

Temporal bone mastoid region; classic location



8) PLASMACYTOMA

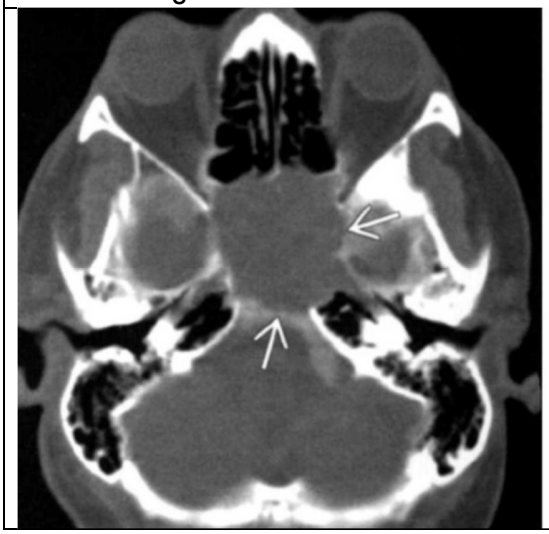
- Multiple myeloma (MM) is a malignant plasma cell disorder that usually affects the bone marrow.
- 10% of hematologic malignancies.
- Ages : 40 – 80y
- Sex: M > F

Location:

- Sino nasal region = most common location in head &neck
- Skull base (especially sphenoid body, petrous temporal bone)
- Calvarial marrow space

Imaging findings:

- Solitary intraosseous osteolytic soft tissue mass with non-sclerotic margins.



Intra-Cranial Cystic Lesions

Extra-axial	Intra-axial Parenchymal	Intra-ventricular
• Arachnoid cyst • Dermoid • Epidermoid • Pineal cyst • Rathke cleft cyst • Cystic tumors: Craniopharyngioma Schwannoma	• Enlarged PVS = perivascular spaces • Neuroglial cyst • Porencephalic cyst • Choroidal fissural cyst • Cystic tumors: 1) Pilocytic astrocytoma 2) Hemangioblastoma 3) Ganglioglioma 4) DNET 5) PXA 6) Cystic Metastasis	• Cavum septum pellucidium *(CSP)* • Cavum vergae *(CV)* • Cavum veli interpositi *(CVI)* • Choroid plexus cyst • Ependymal cyst • Colloid cyst • Arachnoid cyst • Trapped ventricle by tumor

Intra-axial Parenchymal Cysts

1) Enlarged PVSs = Virchow- Robin Spaces

- Pial-lined interstitial fluid-filled structures that accompany penetrating arteries and veins.
- They do not communicate directly with the subarachnoid space.
- Common, incidental, “leave me alone” lesions.

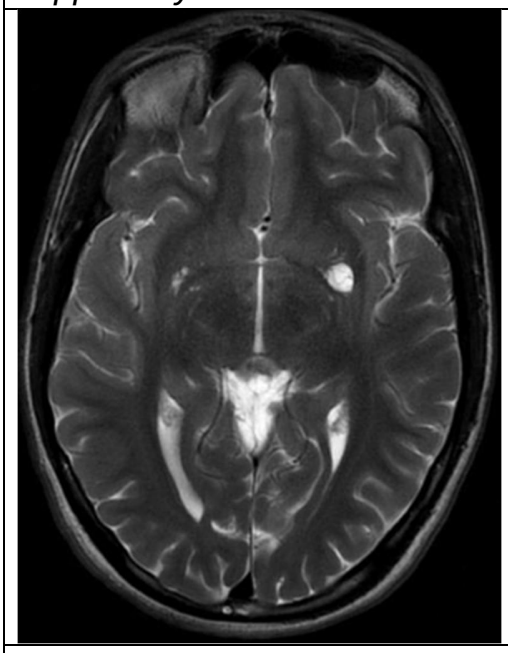
<u>Common Location:</u>

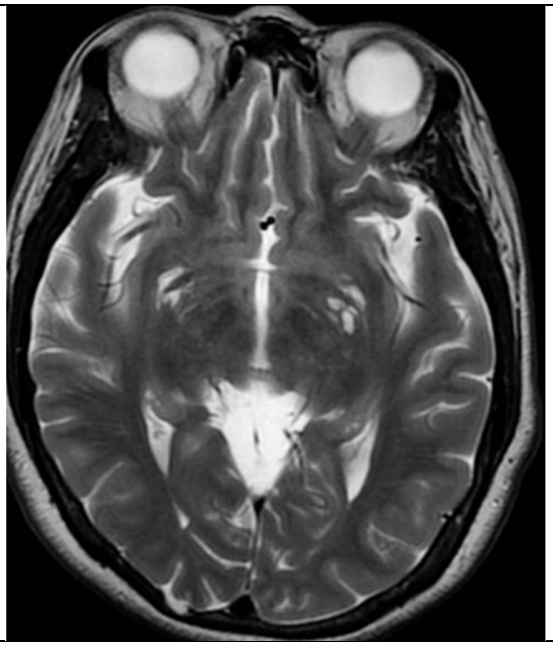
- Inferior basal ganglia, clustering around the anterior commissure and surrounding the lenticulostriate arteries

<u>Other common locations:</u>

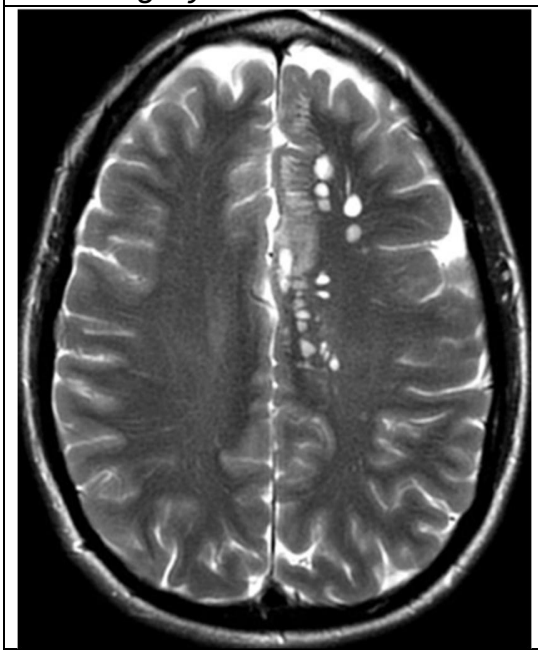
- Midbrain, deep white matter, and subinsular cortex.
- Thalami, dentate nuclei, corpus callosum, and cingulate gyrus

Type 1: VRS in the inferior basal ganglia especially in the anterior perforated substance along the line of the anterior commissure Supplied by lenticulostriate arteries

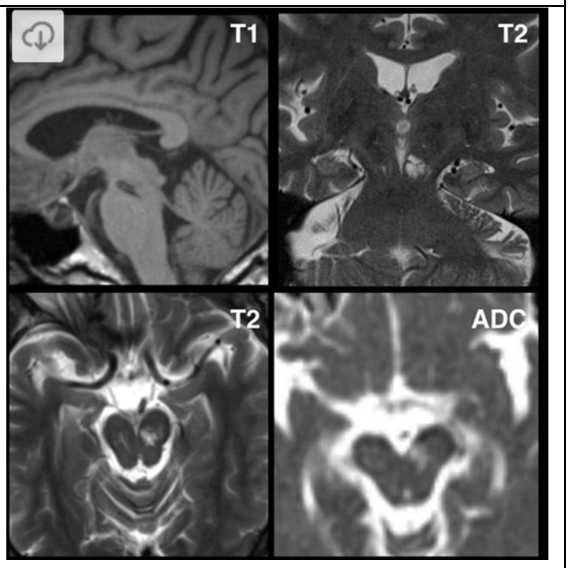




Type 2: VRS in the deep WM supplied by the perforating medullary arteries as they enter the cortical gray matter



Type 3: VRS in the midbrain



2) NEUROGLIAL CYSTS

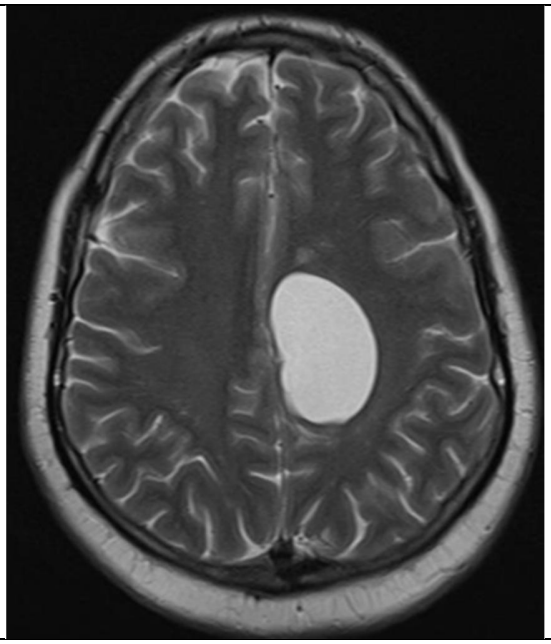
- Congenital lesions, arising from embryonic neural tube elements that become sequestered within the developing white matter.
- Rare benign cystic lesion

Location:

- 1st common location: Frontal lobe
- 2nd most common location: Temporal lobe

Imaging findings:

- Unilocular cystic lesion
- CT: CSF like density
- MRI: CSF like signal intensity
- +C: No enhancement
- No calcification
- No surrounding edema
- No mass effect

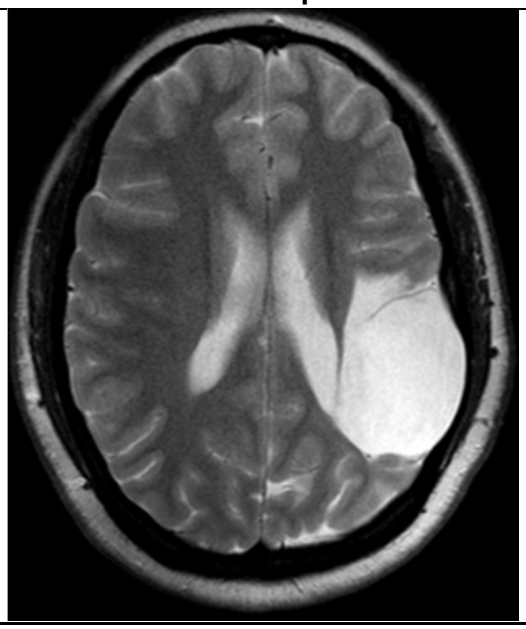
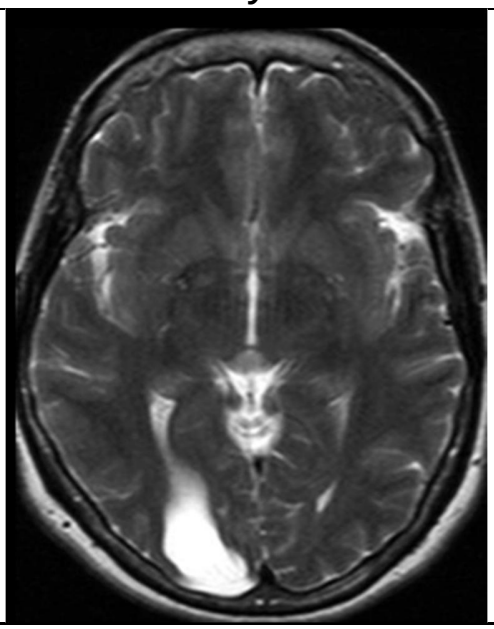
Axial CT	Axial T2
	

3) Porencephalic Cyst

- Porencephaly is a rare congenital disorder.
- Results from encephaloclastic insult (e.g. intrauterine infections, ischaemia and hemorrhage) => Cystic degeneration and encephalomalacia

Imaging findings = CSF like signal:

- Intaraparenchymal well defined cyst
- Cyst communicating with ventricular system and / or subarachnoid space
- Cyst lined by white matter *(NOT GM DD: Schizencephaly)*
- CT: hypodense = CSF attenuation
- T1: low signal intensity
- T2: high signal intensity
- FLAIR: suppression of fluid signal intensity
- DWI: no restricted diffusion

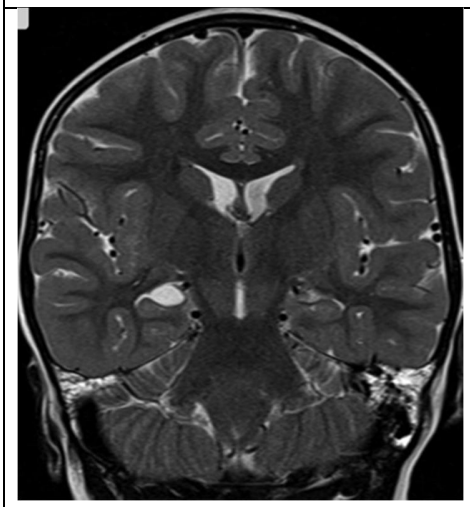
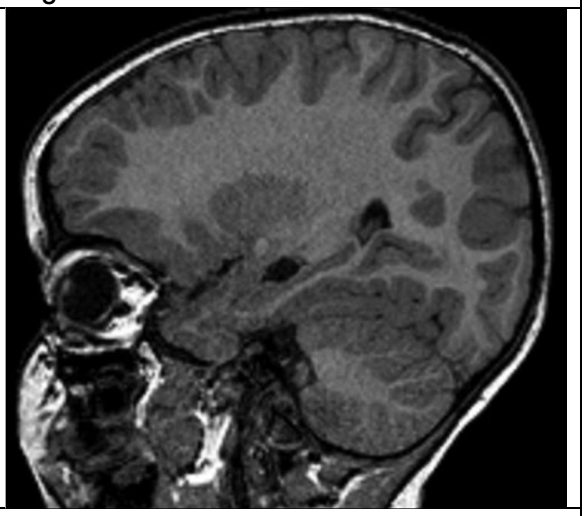
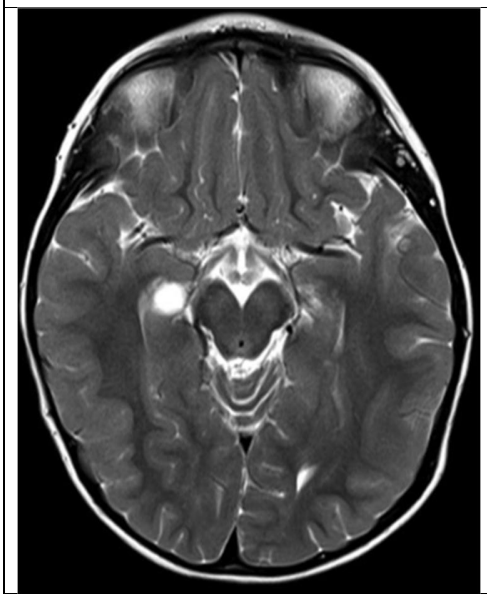
Communicated with subarachnoid space	Communicated with ventricular system
	

4) Choroidal Fissure Cyst

- A benign intracranial cyst occurring at the level of choroidal fissure

Imaging findings:

- A well defined intracranial spindle or ovoid shaped cyst
- Paralleling the long axis of the temporal lobe and choroid fissure on sagittal images
- CT = CSF density
- MRI = CSF like signal
- No calcification, surrounding edema, mass effect, enhancement.

Coronal T2	Sagittal T1
	
Axial T2	**Axial T1+C**